Primær Skleroserende Cholangitt

Lise Katrine Engesæter
Lege/stipendiat
OUS Rikshospitalet

NO PSC
Norwegian PSC Research Center

Oversikt

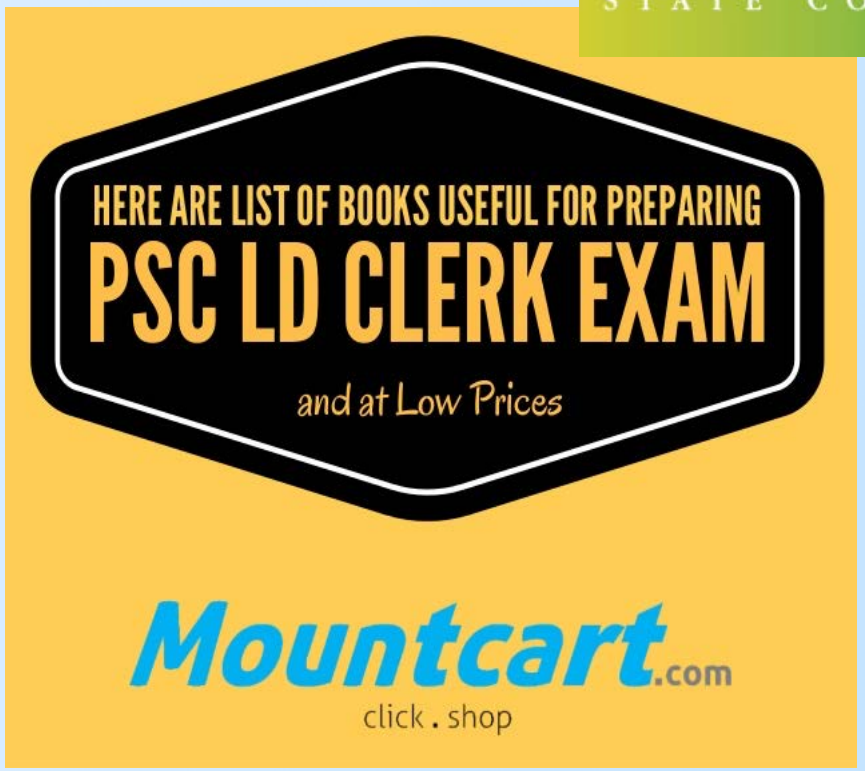
- Hva er PSC
- Diagnostikk
- Varianter av PSC
- PSC; tarmbetennelse og kreft
- Oppfølging & behandling →Nye retningslinjer
- Oppsummering

PSC
Est. 1960
Plymouth
Swim Club

PSC
PENSACOLA
STATE COLLEGE

PETITCODIAC
SPORTSMAN'S CLUB

SPeD
PSC
NCCA


HERE ARE LIST OF BOOKS USEFUL FOR PREPARING
PSC LD CLERK EXAM
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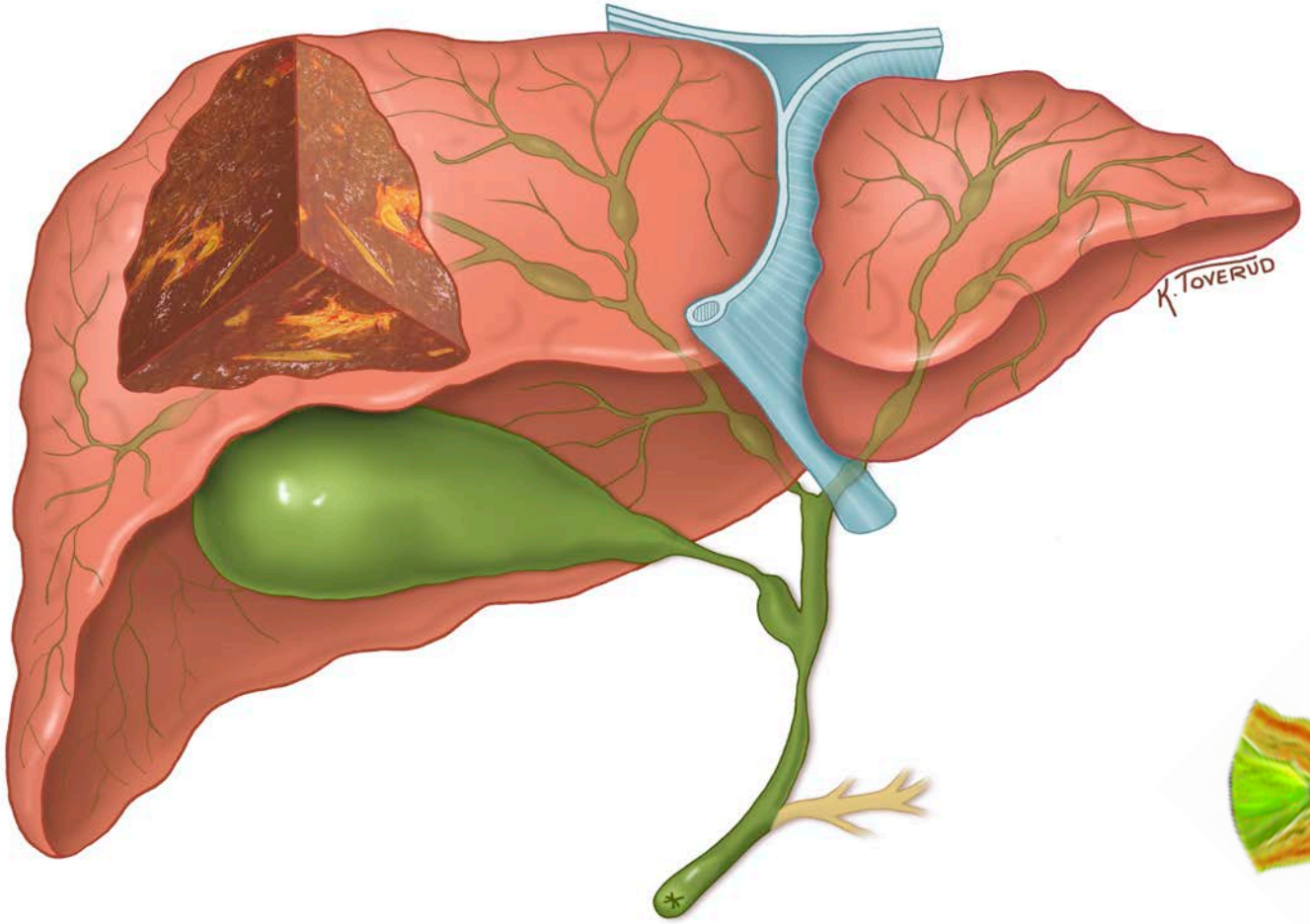
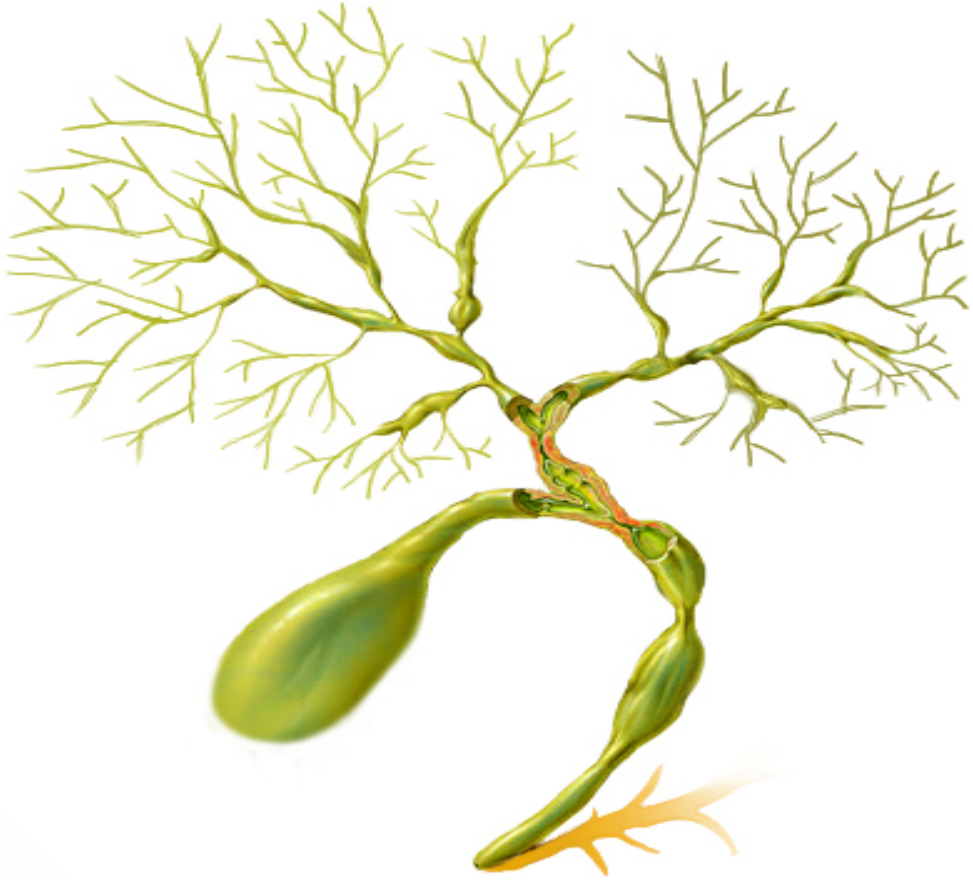
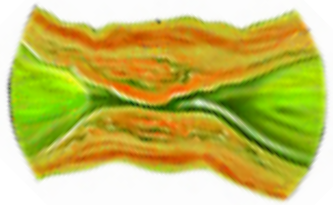
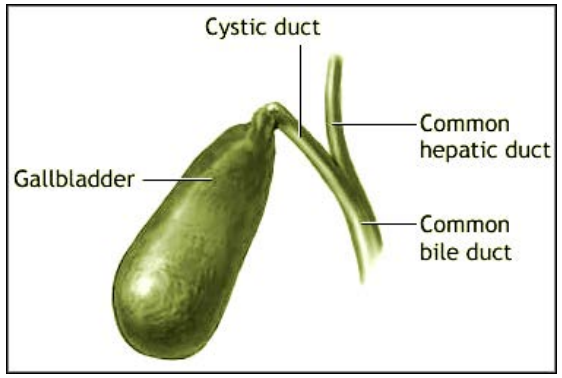
KERALA PSC
GOVERNMENT OF KERALA
सत्मेव जयते
EXAMS

Primær	Ukjent årsak
Skleroserende	Arrete og stivt/trangt
Cholangitt	Betennelse i galleveiene


K. Toverud

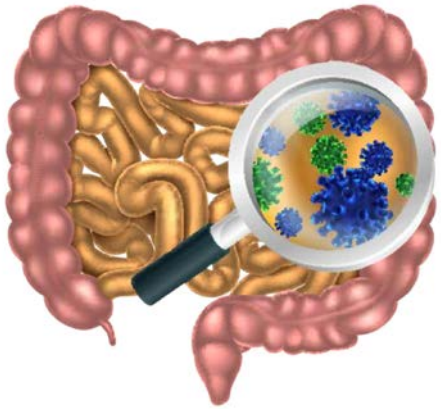






→Galle

→Autoimmun sykdom



→Tarmflora

Hvordan stilles diagnosen?

- Utfordrende diagnostikk:
  - Pasienter uten symptomer
  - Blodprøver kan være varierende
- ERC(P) eller MRC(P) er diagnostisk

- Leverbiopsi ikke nødvendig for diagnose
- Sekundær skleroserende cholangitt må utelukkes.

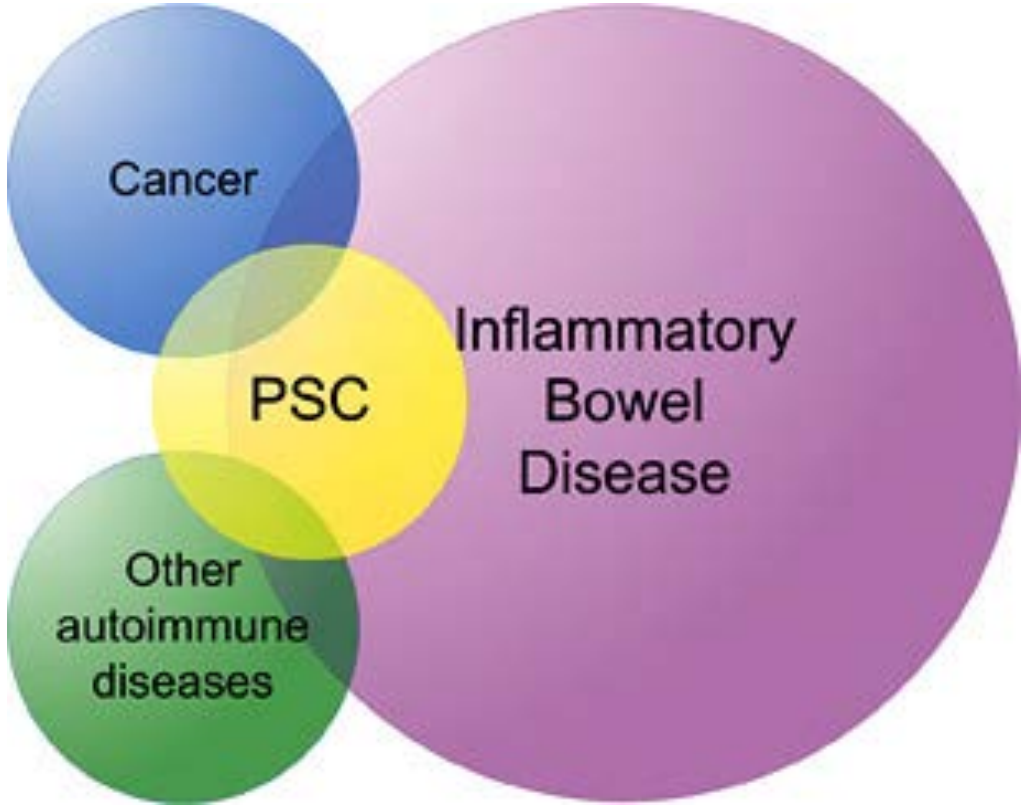
Spesielle varianter av PSC

Small-duct PSC

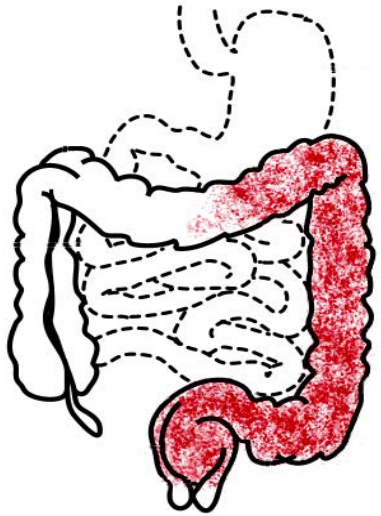
- 6-16%
- Normal galletre på MR/ERCP
- Samme symptomer
- «snillere» forløp enn vanlig PSC

PSC/AIH

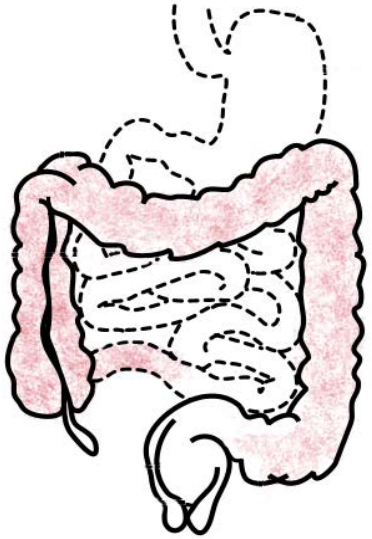
- 7-14%
- AIH synes ikke på MR
- Før beskrevet som «overlapp»



- Ca 25% har annen autoimmun sykdom
- Ca 80% har IBD(tarmbetennelse)
  - Usikker sammenheng
  - Oftere betennelse i hele tarmen
  - IBD og PSC utvikles uavhengig av hverandre
  - IBD kan ha et helt fredelig forløp



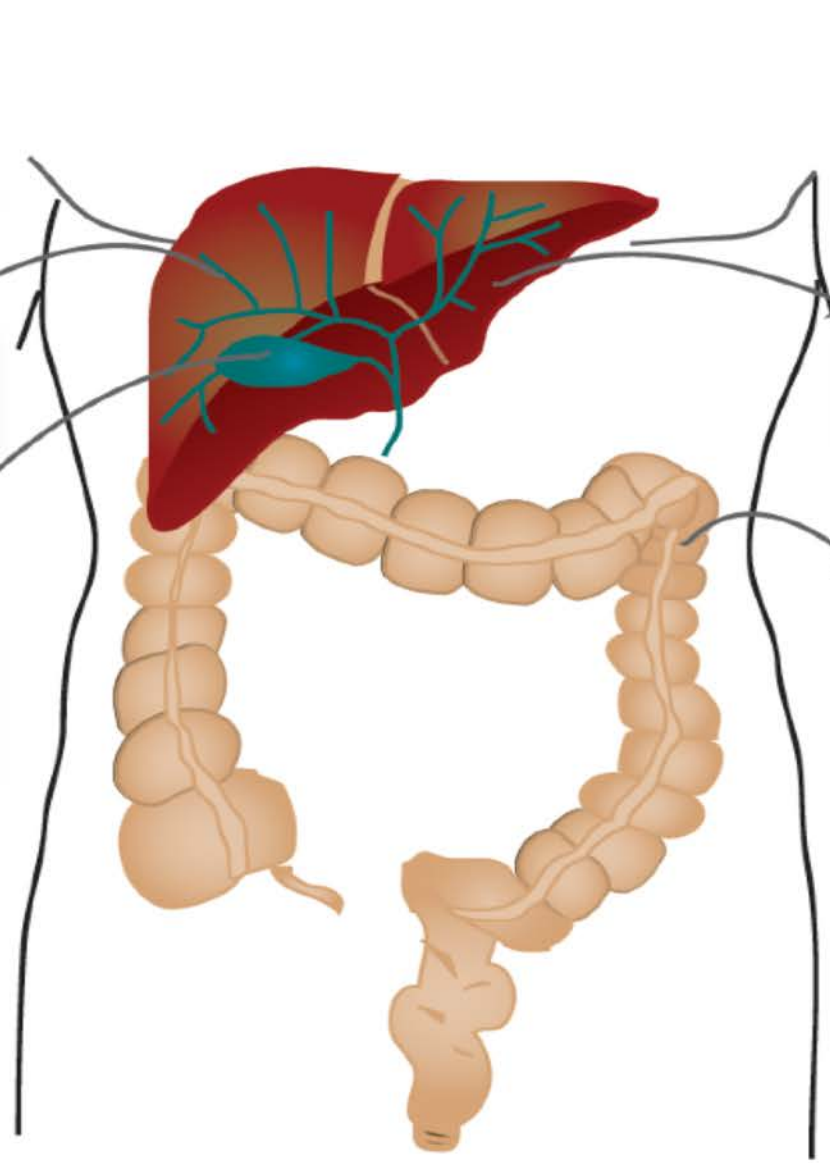
IBD



PSC-IBD

Galleblærekreft hos 2-4%

Gallegangskreft hos 7-14%

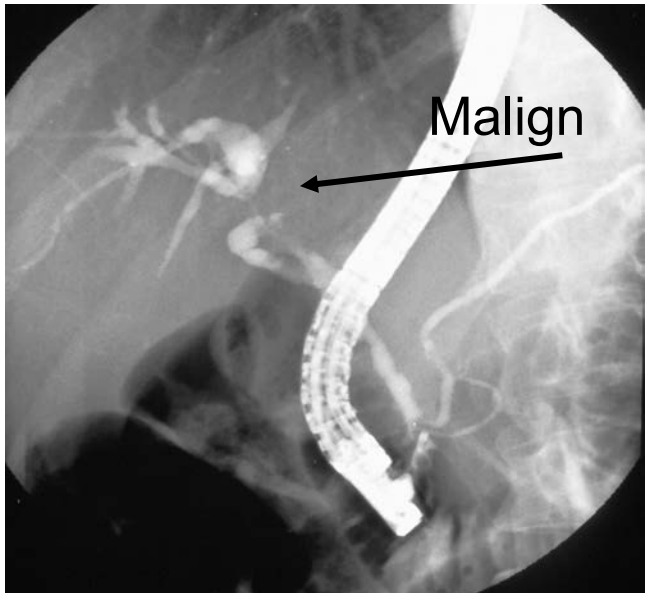


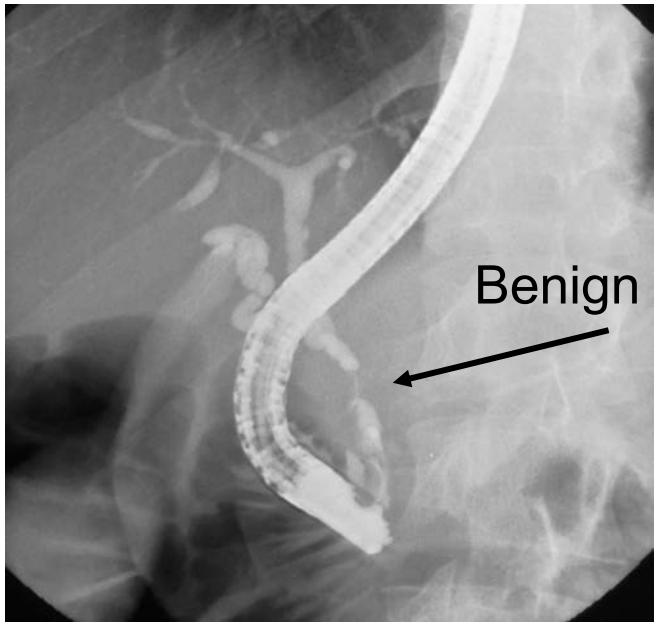
Leverkreft hos 2-4%

Tykktarmskreft ;
15% risiko etter 10 år, 30% risiko etter 20 år

Med utgangspunkt i Boberg K.M., Lind G.,Best Pract Res Clin Gastroenterol , 2011

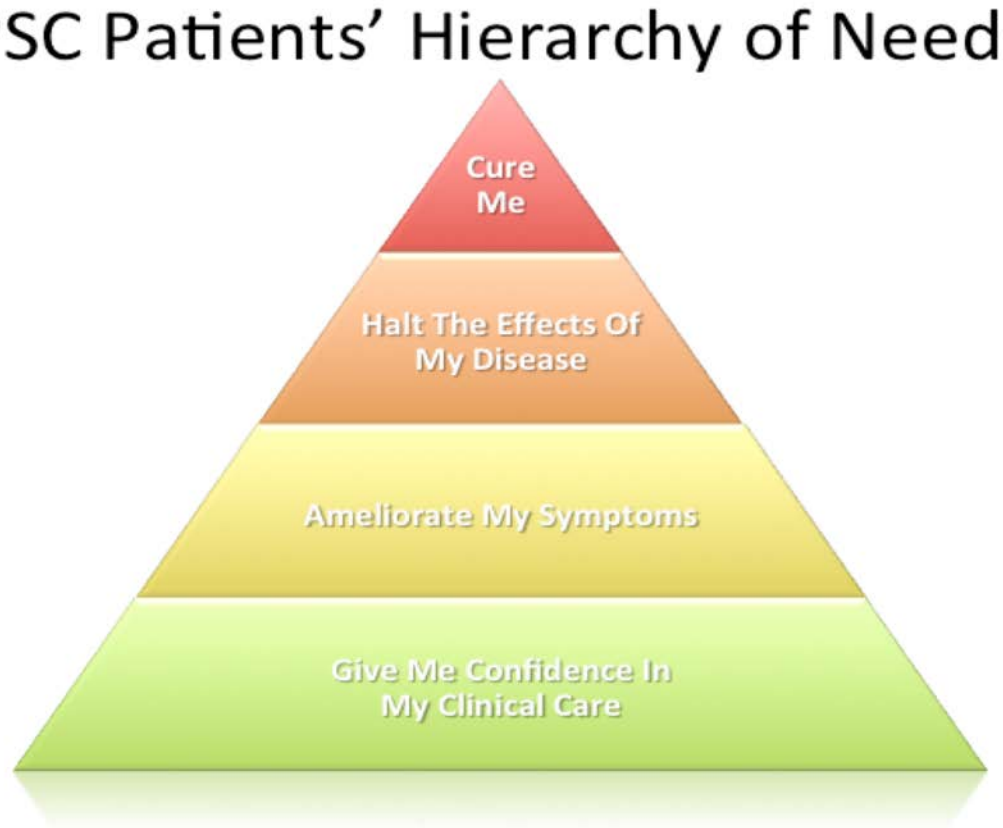
- 7-14% livstidsrisiko
- 30-50 % diagnostiseres < 1 år fra diagnose
- Utredning bør skje i tverrfaglig team
- Vanskelig diagnose
  - CT, MR
  - Ca19-9
  - Børstecytologi(celleprøve fra gallegangen)
- Langtidsoverlevelse ved tidlig diagnose og kirurgi





Behandling

- Medisiner
- Endoskopi
- Levertransplantasjon



https://www.pscsupport.org.uk/sites/all/themes/psc/logo.png

Should people with PSC be treated with ursodeoxycholic acid?

Recommendations

- UDCA at doses of 15-20 mg/kg/d can be given since it may improve serum liver tests and surrogate markers of prognosis. Available data does not allow for a firmer recommendation **(LoE 1, weak recommendation, 76% consensus).**
- UDCA at doses of 28-30 mg/kg/d should not be given **(LoE 1, strong recommendation, 100% consensus).**

Ursodeoksykolsyre(UDCA) *kan* gis i mindre doser siden det forbedrer noen leverprøver.

Should people with PSC be treated with long-term antibiotics to prevent disease progression or decrease PSC-related complications?

Recommendation

- Long-term use of antibiotics is not recommended for treatment of PSC in the absence of recurrent bacterial cholangitis **(LoE 3, strong recommendation, 100% consensus).**

Langtidsbehandling med antibiotika er ikke anbefalt.

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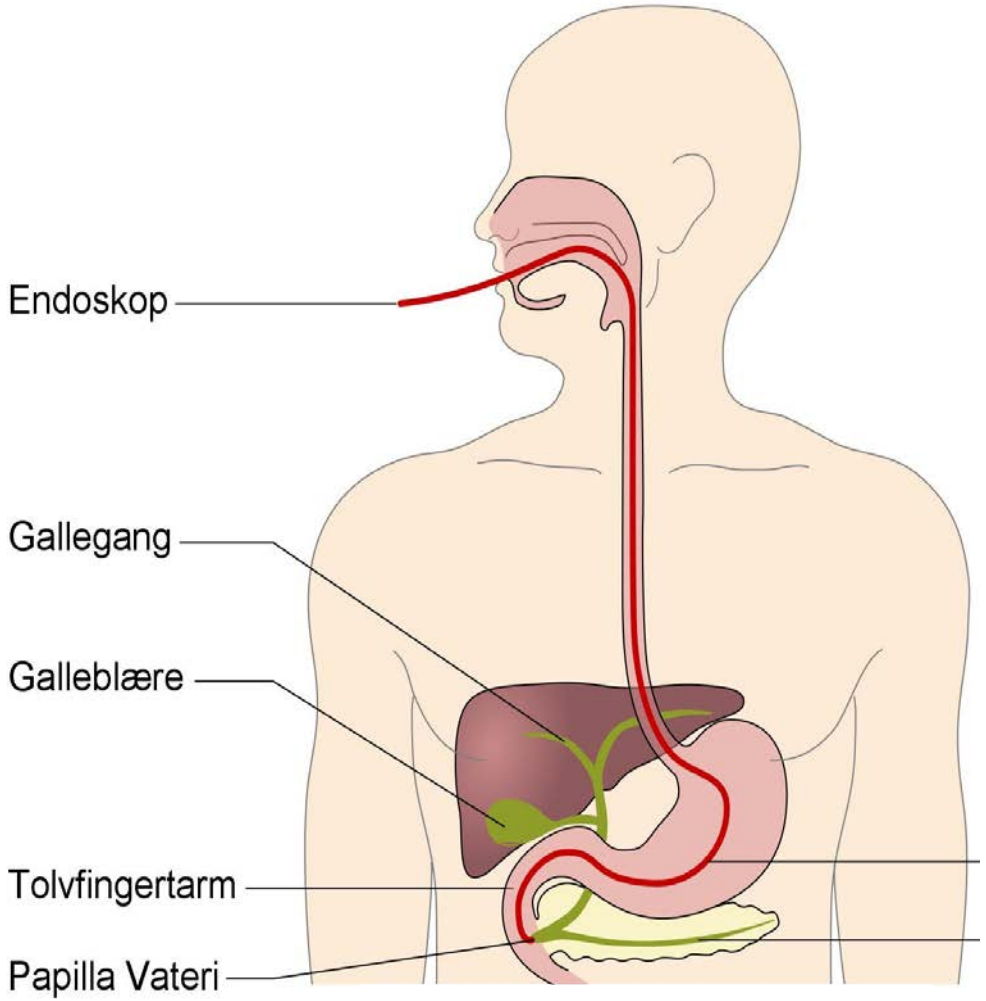
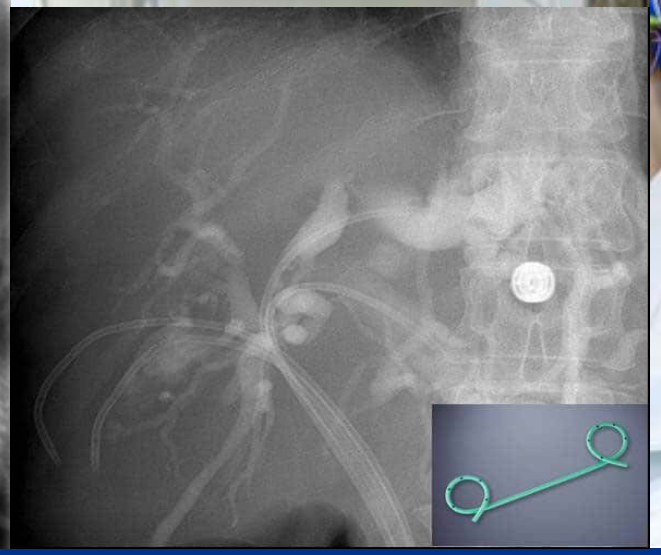
Endoskopi(ERCP)

- Ofte i etterkant av MR
- Ballongdilatering
- Mulighet for celleprøver
- Evt stent for kortere perioder
- Fare for komplikasjoner



ERCP på Rikshospitalet



Levertransplantasjon

- Rikshospitalet siden 1984
- Ca 100 operasjoner per år
- PSC, leverkreft, alkoholisk leversykdom vanligste indikasjoner
- Pasienter med PSC har god prognose
- Kraftig utvikling
  - 1991 → 9 timer 35 minutter og 20 poser blod
  - I dag→ 5 timer 35 minutter and 0-1 poser blod

Foto:Rune Thorstein,Dagens Medisin

	1982-90	1991-96	1997-01	2002-06	2007-11	2012-16	2017-21	2021
Alcoholic cirrhosis	1.9 %	8.4 %	12.2 %	12.6 %	11.3 %	11.4 %	15.0 %	16.2 %
Primary sclerosing cholangitis	11.1 %	12.9 %	15.6 %	14.6 %	17.3 %	16.1 %	16.4 %	15.2 %
Hepatocellular carcinoma and cirrhosis	10.8 %	4.4 %	6.4 %	7.8 %	11.3 %	16.8 %	15.2 %	12.9 %
Metabolic disease	9.3 %	7.5 %	5.9 %	5.7 %	7.0 %	7.8 %	9.5 %	9.6 %
Cirrhosis - unknown	0.6 %	3.1 %	3.0 %	3.4 %	5.5 %	6.6 %	6.0 %	8.4 %
Acute liver failure - other	8.7 %	10.0 %	7.7 %	6.3 %	5.4 %	5.2 %	6.5 %	6.3 %
Polycystic disease	0.3 %	1.2 %	0.9 %	1.7 %	1.4 %	2.1 %	3.0 %	5.1 %
Autoimmune cirrhosis	2.8 %	4.1 %	2.8 %	4.4 %	4.4 %	4.7 %	4.4 %	3.8 %
Secondary liver tumors	0.9 %	0.5 %	0.8 %	0.8 %	2.2 %	1.8 %	2.2 %	3.5 %
Primary biliary cirrhosis	22.6 %	13.6 %	7.9 %	7.4 %	6.0 %	5.3 %	4.2 %	2.5 %
Post hepatitis C cirrhosis	0.0%	3.7 %	8.2 %	10.6 %	9.7 %	7.4 %	2.4 %	2.5 %
Acute liver failure - toxic	0.6 %	2.8 %	4.4 %	4.1 %	3.3 %	2.9 %	2.6 %	2.3 %
Extrahepatic biliary atresia	6.8 %	5.4 %	4.5 %	4.5 %	2.4 %	2.4 %	2.6 %	1.3 %
Other liver malignancies	2.5 %	2.3 %	1.8 %	1.5 %	1.2 %	1.3 %	1.0 %	1.3 %
Cholangiocarcinoma	1.5 %	0.9 %	0.8 %	0.7 %	0.6 %	0.2 %	0.7 %	1.0 %
Other	18.9 %	19.1 %	17.0 %	13.9 %	10.8 %	8.0 %	8.0 %	6.3 %

NLTR Annual report 2021 prepared by Espen Melum

When should liver transplantation be considered in people with PSC?

Recommendations

- Liver transplantation should be considered for people with PSC and decompensated cirrhosis or hepatocellular carcinoma according to standard guidelines **(LoE 3, strong recommendation, 100% consensus).**

- Liver transplantation should be considered for people with PSC with recurrent bacterial cholangitis and/or severe pruritus or jaundice despite endoscopic and pharmacological therapy **(LoE 3, strong recommendation, 100% consensus).**
- Liver transplantation can be considered in people with PSC and high-grade biliary dysplasia confirmed by cytology or ductal histology **(LoE 4, weak recommendation, 92% consensus).**
- Liver transplantation for early-stage CCA in PSC can be performed within the context of clinical trials **(LoE 4, weak recommendation, 92% consensus).**

- Skrumplever/Leversvikt
- Leverkreft
- Tilbakevendende infeksjoner
- Kløe
- Celleforandringer
- Gallegangskreft “hvis del av forskning”

Box 3. Suggested algorithm for follow-up of primary sclerosing cholangitis.

A) Routine surveillance (reappraisal of disease progression and risk)

- Every 12 months (for all, every 6 months in patients with significant risk):
  - Clinical evaluation (including quality of life)
  - Serum liver-related tests including bilirubin, ALP, AST, platelets, and PT
- Every 12 months (even for patients at low risk):
  - MRI/MRCP and/or US (with a special attention for gallbladder wall abnormalities)* §
  - Colonoscopy**
  - Elastography and/or ELF test
- Every 2 to 4 years (for all): DEXA for bone mineral density assessment (and serum vitamin D measurement)

B) Additional work-up when clinically indicated (new symptoms or evolving abnormalities in routine investigations (ALP/bilirubin rising) or ΔLSM >1.5 kPa/year or ductal progression):

- Suspected cholangiocarcinoma: serum CA 19.9 and MRCP/MRI liver with contrast and ERCP with cytologic or histologic sampling
- Suspected features of auto-immune hepatitis or drug toxicity: serum IgG and autoantibodies of AIH, consider liver biopsy
- Suspected clinically relevant portal hypertension (Baveno VII criteria[114]): EGD, consider non-selective beta blockers

In UDCA treated patients, consider non-compliance

Rutinekontroller hver 12.mnd

- Klinisk vurdering
- Blodprøver
- MR og/eller ultralyd
- Koloskopi
- Sjekk for leverfibrose

Benmineralmåling hvert 2. til 4.år

"Ekstrakontroller"

- Ca 19-9
- MR med kontrast
- ERCP
- Autoimmun hepaitt?

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Recommendations

- CCA must be suspected in i) newly diagnosed PSC with high-grade stricture(s) and in ii) known PSC with worsening of signs or symptoms, progressive stricture(s) or a new mass lesion identified on imaging **(LoE 4, strong recommendation, 93% consensus).**
- Diagnostic work-up by an experienced multidisciplinary team is recommended in people with PSC and suspected CCA **(LoE 5, strong recommendation, 100% consensus).**
- Contrast-enhanced, cross-sectional imaging is recommended as the initial diagnostic test when CCA is suspected, potentially followed by ERCP with ductal sampling (brush cytology, endobiliary biopsies) for diagnosis and staging of the suspected CCA **(LoE 1, strong recommendation, 96% consensus).**
- Serum CA 19-9 can be assessed in all patients where CCA is suspected and fluorescence *in situ* hybridisation (FISH) or equivalent chromosomal assessments can be considered when brush cytology and/or histology are equivocal **(LoE 3, weak recommendation, 91% consensus).**

Mistenk gallegangskreft ved:

1. Ny PSC diagnose med mye gallegangsstrikturer
2. Kjent PSC med forverring

Oppfølging av erfarent tverrfaglig team
ved mistanke om gallegangskreft

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When should people with PSC be referred to an experienced centre in PSC care?

Recommendation

- Initial expert consultation for people with PSC at diagnosis and referral for those with symptomatic and/or progressive PSC to an experienced centre with ready access to PSC clinical trials and a dedicated multidisciplinary team are recommended **(LoE 5, strong recommendation, 100% consensus).**

1. Ved diagnose
2. Ved forverring

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What should be the role of patient support groups?

Recommendation

- Information on existing patient support groups should be provided (**LoE 5, strong recommendation, 100% consensus**).

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Hva bør være forskningsfokus fremover?

- Verktøy for å stille diagnosen PSC tidlig
- Verktøy for å oppdage gallegangskreft tidlig
- Avklare hva som er gode mål når man tester nye medisiner
- Finne årsaken til PSC
- Starte kliniske studier

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Hva skal man huske om PSC?

- Variert forløp
- MR viktigst for å sette diagnosen
- Ingen kur, men allikevel behandling
- PSC er god indikasjon for levertransplantasjon
- PSC pasienter skal ha systematisk oppfølging

Clinical Practice Guidelines

JOURNAL OF HEPATOLOGY

EASL Clinical Practice Guidelines on sclerosing cholangitis☆

European Association for the Study of the Liver*

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"You have an extremely, rare, hard-to-treat disease — are you trying to make me look bad?"

Spørsmål?