Primær skleroserende cholangitt

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KERALA PSC
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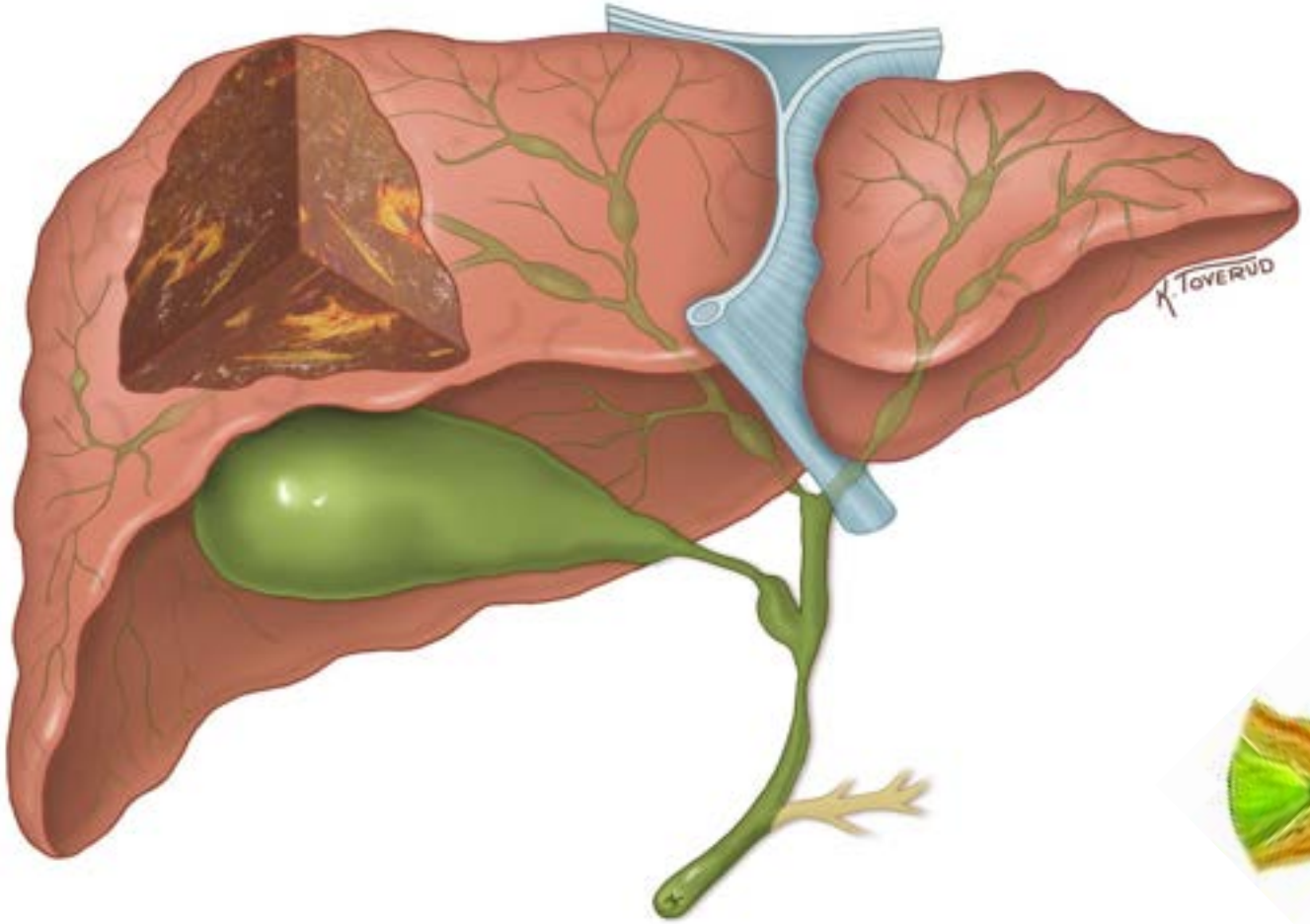
EXAMS

Oversikt

- Hva er PSC
- Diagnostikk
- Varianter av PSC
- Tarmbetennelse og kreft
- Retningslinjer oppfølging & behandling
- Oppsummering

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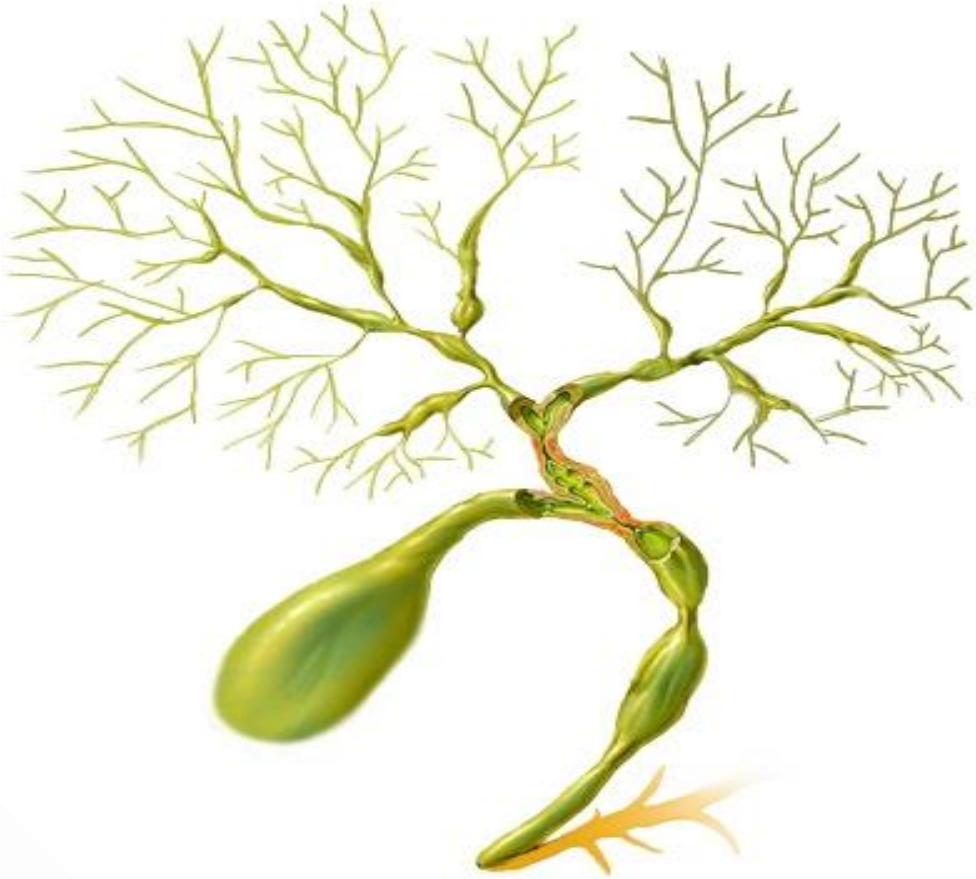
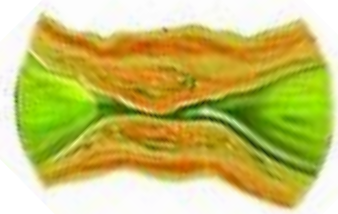
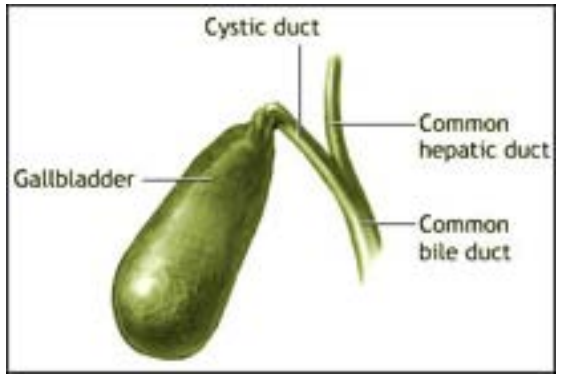
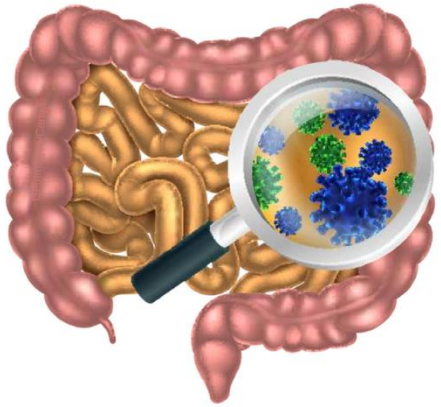






→Galle

→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.

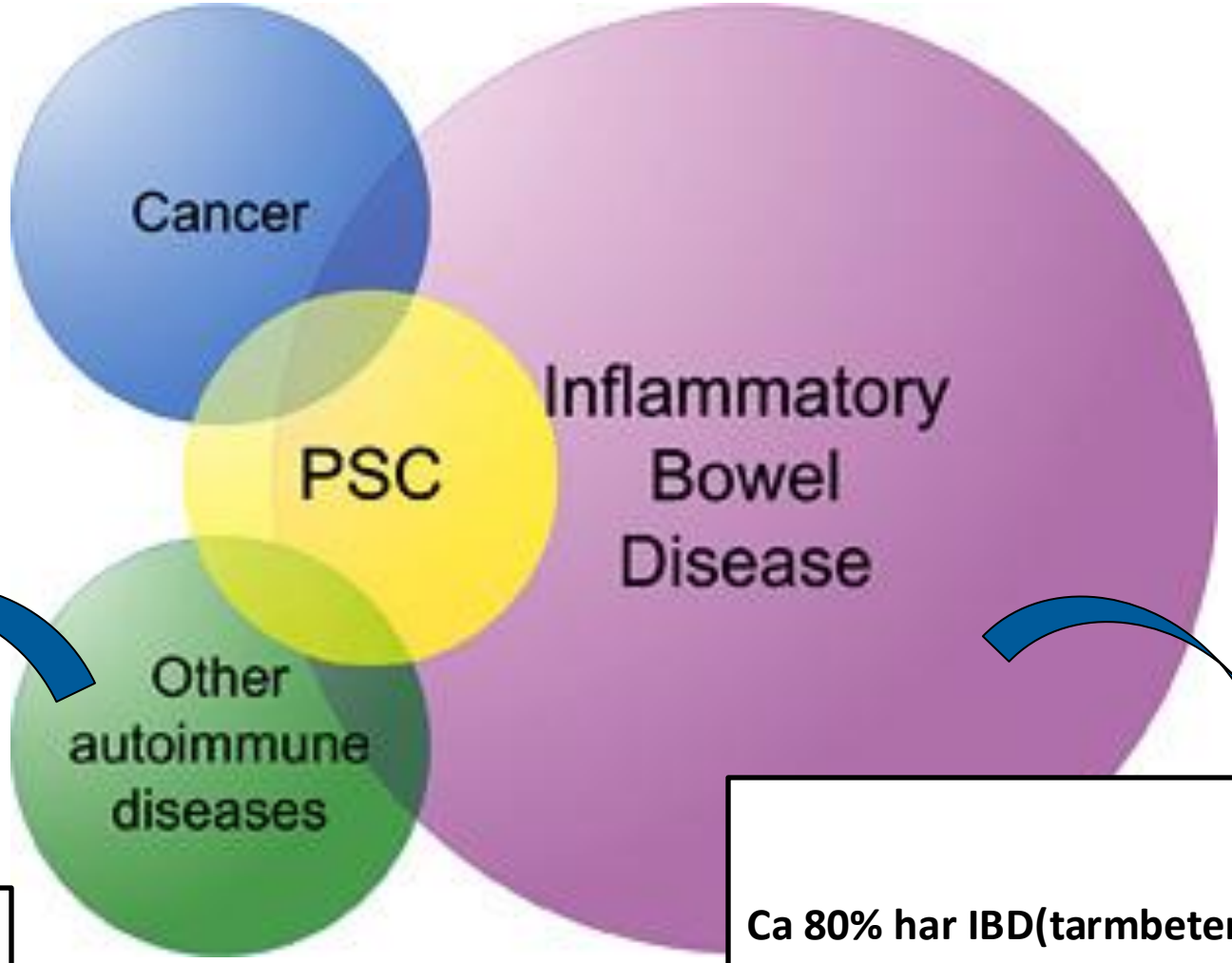
Spesielle varianter av PSC

❖Small-duct PSC

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

❖PSC/AIH

- 7-14%
- Autoimmun hepatitt synes ikke på MR
- AIH kan behandles med medisiner
- Før beskrevet som «overlapp»



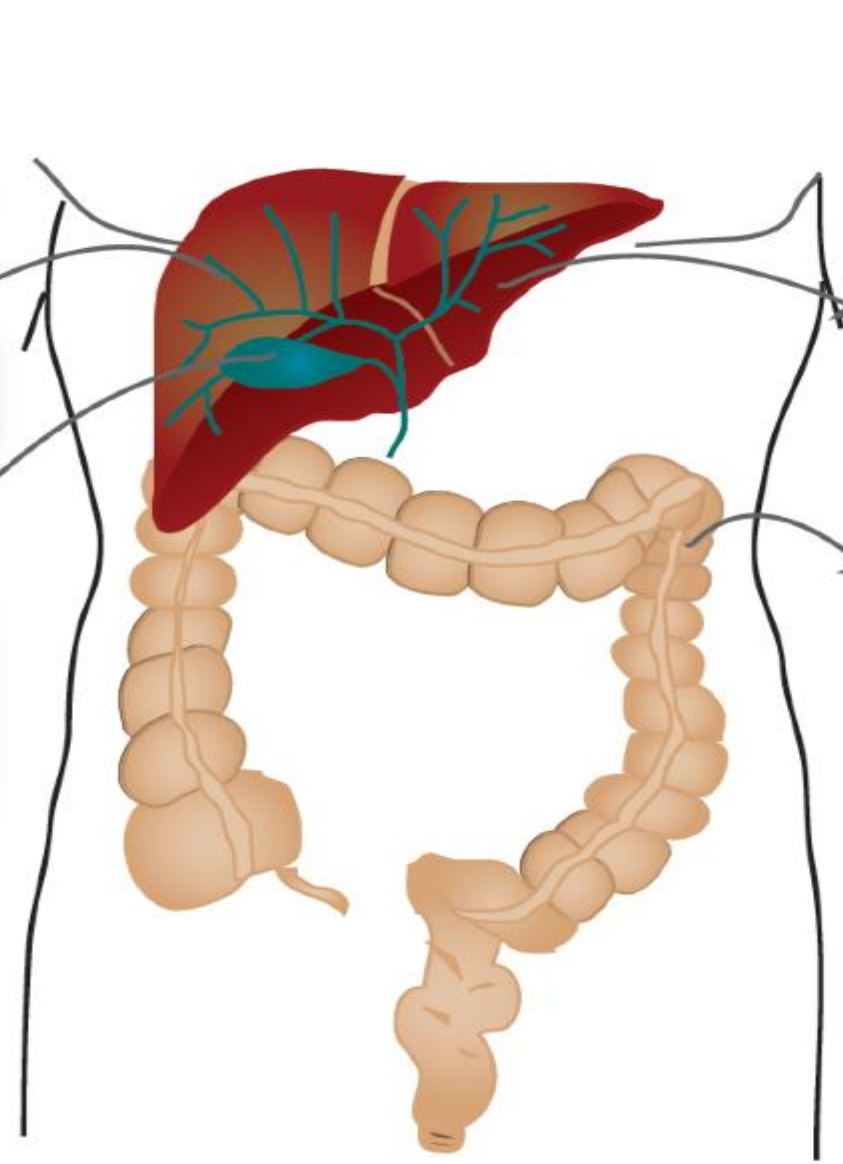
Ca 25 % har annen autoimmun sykdom

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**

Galleblærekreft hos 2-4%

Gallegangskreft hos 7-14%



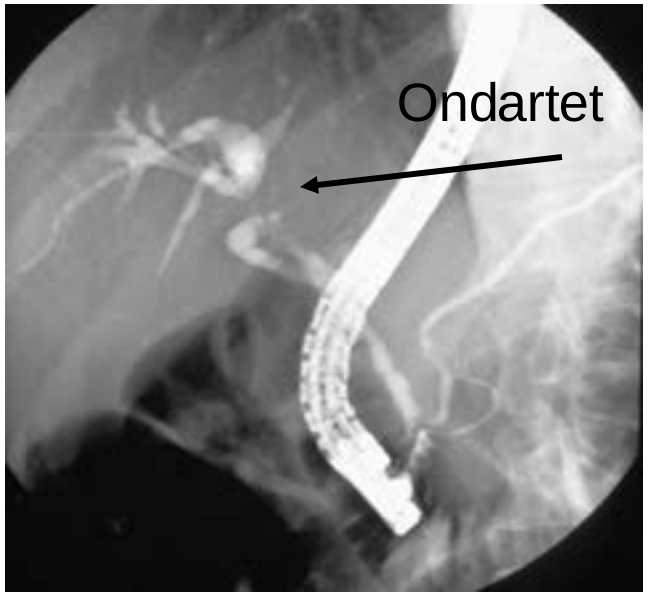
Leverkreft hos 2-4%

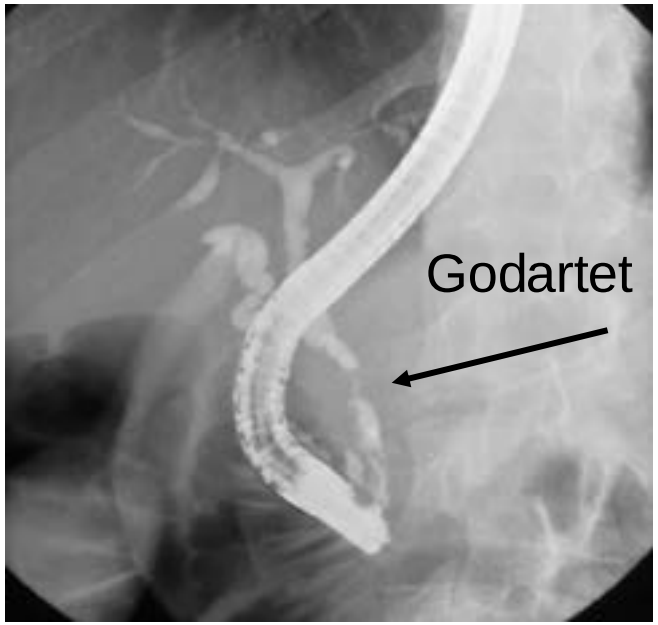
Tykktarmskreft ;
15% risiko etter 10 år,
30% risiko etter 20 år

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

Gallegangkreft(cholangiocarcinom)

- 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





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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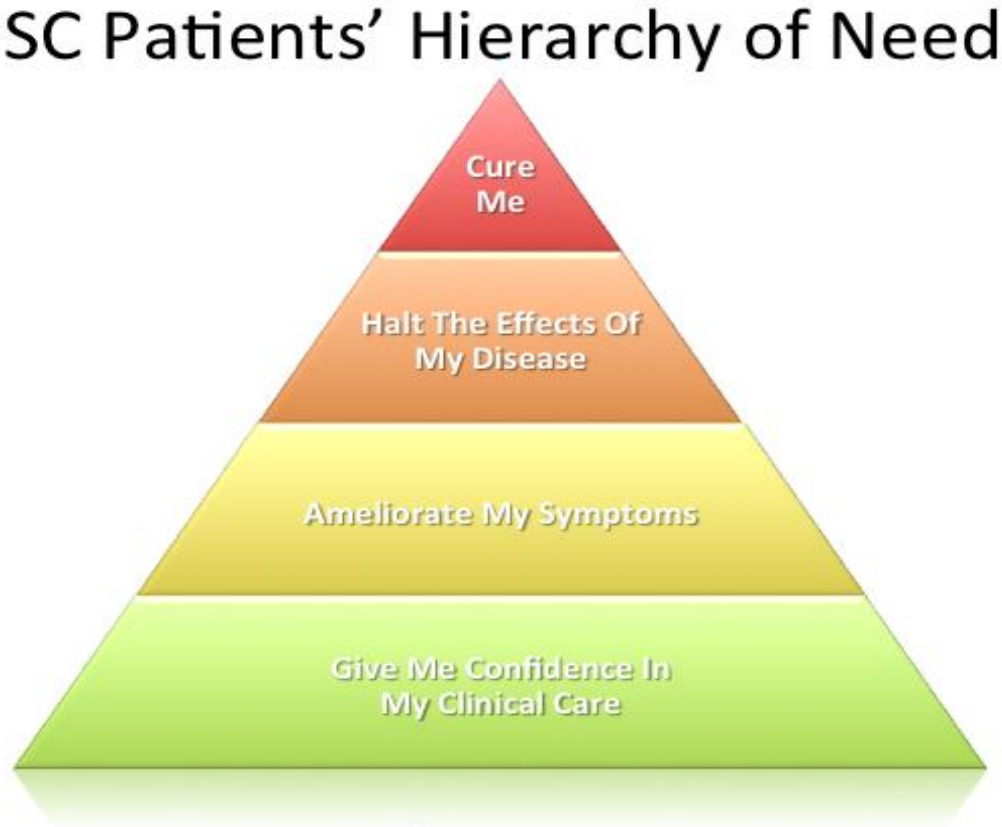
Behandling

I. Medisiner
II. Endoskopi
III. 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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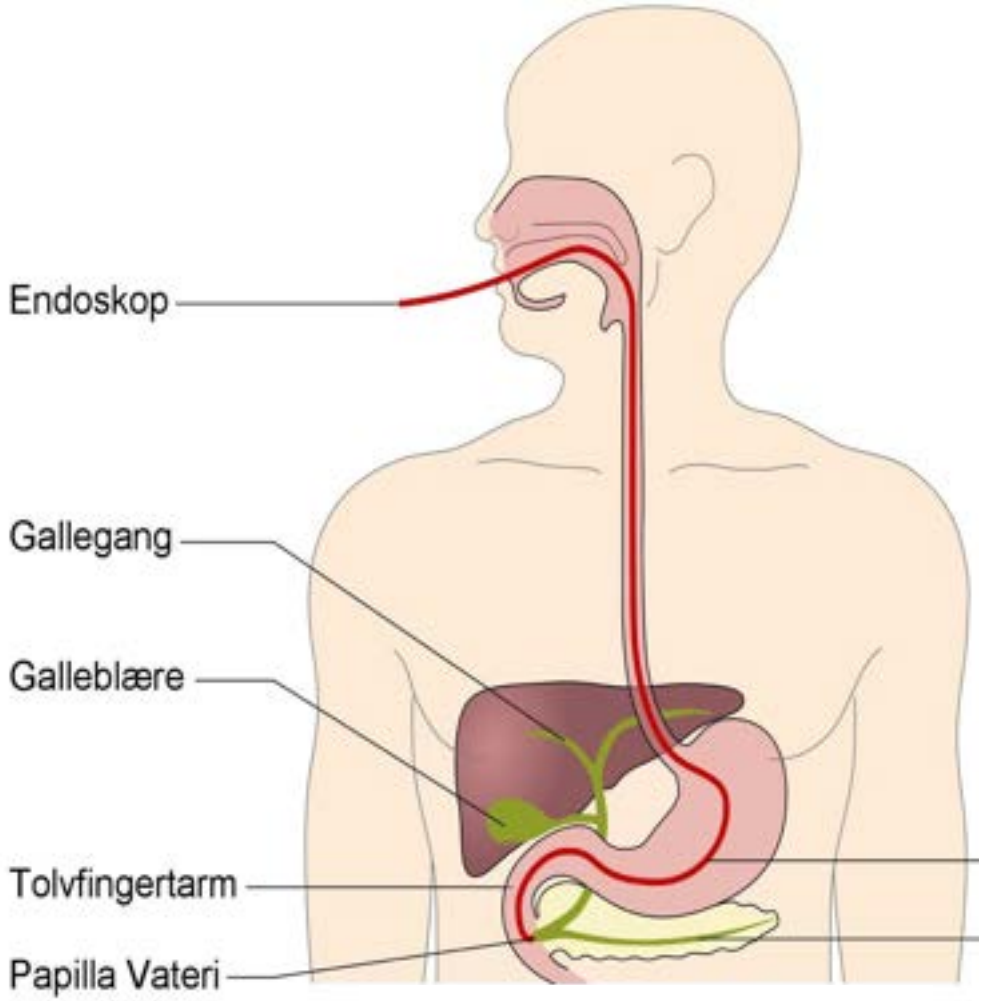
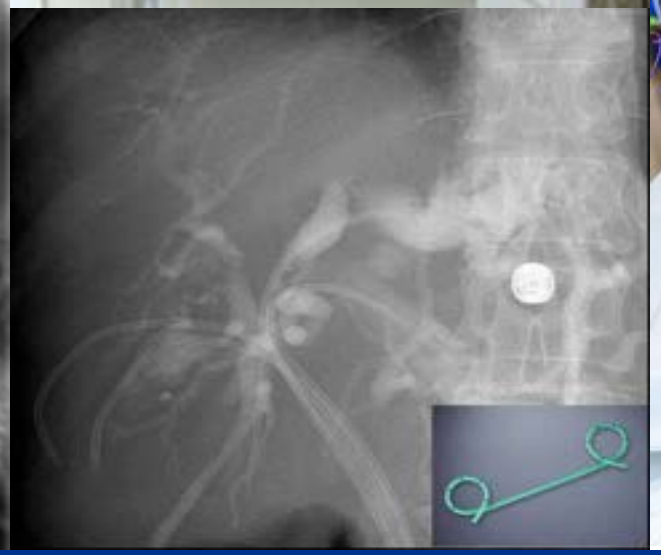
Endoskopi(ERCP)

- Ofte i etterkant av MR
- Ballongdilatering
- Mulighet for celleprøver
- Evt stent for kortere perioder
- Komplikasjoner forekommer



ERCP på Rikshospitalet



Levertransplantasjon

- Rikshospitalet siden 1984
- Ca 90 operasjoner per år
- PSC, leverkreft, alkoholrelatert leversykdom vanligste årsak
- Pasienter med PSC har god prognose
- Kraftig utvikling
  - 1991 → 9 timer 35 minutter og 20 poser blod
  - I dag→ ca 5 timer og 0-1 poser blod

Foto:Rune Thorstein,Dagens Medisin

	1982-90	1991-98	1999-03	2004-08	2009-13	2014-18	2019-23	2023
Hepatocellular carcinoma	10.8%	5.0%	5.6%	9.0%	14.5%	17.2%	14.8%	**17.0%**
Alcohol related liver disease	1.9%	9.9%	11.7%	11.7%	11.5%	12.5%	16.0%	**16.7%**
Primary sclerosing cholangitis	11.1%	13.5%	16.1%	15.7%	14.8%	17.8%	15.7%	**14.8%**
Metabolic disease	9.3%	7.2%	5.6%	5.8%	7.7%	8.4%	9.1%	**9.3%**
Acute liver failure - other	8.7%	9.6%	6.8%	5.8%	5.0%	5.9%	6.1%	**5.0%**
Primary biliary cholangitis	22.6%	12.4%	7.1%	7.4%	5.7%	4.7%	4.2%	**4.8%**
Cirrhosis - unknown	0.6%	3.1%	2.6%	4.5%	6.1%	5.5%	6.7%	**4.3%**
Polycystic disease	0.3%	1.2%	1.3%	1.6%	1.5%	2.5%	3.1%	**4.3%**
Autoimmune cirrhosis	2.8%	3.6%	3.8%	4.7%	4.1%	4.6%	4.3%	**2.6%**
Acute liver failure - toxic	0.6%	2.9%	5.0%	4.1%	3.0%	2.5%	2.5%	**2.4%**
Extrahepatic biliary atresia	6.8%	5.0%	4.6%	3.7%	2.3%	2.4%	2.6%	**2.4%**
Cholangiocarcinoma	1.5%	0.8%	1.0%	0.5%	0.4%	0.4%	0.9%	**2.2%**
Post hepatitis C cirrhosis		4.8%	9.2%	10.5%	10.3%	4.1%	2.1%	**2.2%**
Biliary tract carcinoma			0.2%	0.2%	0.9%	0.8%	0.7%	**1.7%**
Secondary liver tumors	0.9%	0.3%	0.5%	1.8%	1.7%	1.9%	2.5%	**1.7%**
Other	22.0%	20.6%	18.9%	12.9%	10.4%	8.7%	8.8%	**8.6%**

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).**

Indikasjoner for transplantasjon:

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

Retningslinjer for oppfølging

Rutinekontroll (årlig)

- Klinisk vurdering og samtale
- Blodprøver
- MR og/eller ultralyd
- Koloskopi
- (Sjekke for leverfibrose)

Ekstrakontroll (ved forverring)

- MR med kontrast
- ERCP
- Ca 19-9
- Vurdere annen leversykdom

Annet (hvert 2.-4.år)

- Benmineralmåling hvert 2.-4. år

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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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Forskningsfokus

- 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(og tilbakefall av PSC)
- Starte kliniske studier

NO PSC
Norwegian PSC Research Center

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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*

Spørsmål?