National audit of diagnosis, surveillance & management of Primary Sclerosing Cholangitis (PSC) in the United Kingdom

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Background & aim

- Primary sclerosing cholangitis (PSC) is a rare disorder and as such clinical care can be heterogeneous.
- We audited PSC management across the UK against audit standards set by the recently published British Society of Gastroenterology (BSG) guidelines on PSC management. (Chapman MH, et.al. Gut. 2019)
- BASL and BSG endorsement

Methods

UK-PSC

- All UK PSC investigators were invited to complete an electronic questionnaire on the PSC patient cohort encompassing

✓Diagnosis

✓Demographics

✓Bowel and biliary tract cancer surveillance

✓Risk stratification assessment for liver disease

- Data were collected between March 2019 and January 2021.

Results

- 1,795 patients were included across 30 UK centres
- Liver units n = 1548
- General gastroenterology units n = 247
- Median age was 51 years
- 56.4 % were men

Fatourou et.al. Gut 2021;**70:**A9

UK-PSC

Liver disease management

Follow up and diagnosis

- Majority of patients were followed up by a hepatologist (n = 1610, 89.7 %).
- Magnetic resonance cholangiography (MRCP) was performed as a diagnostic investigation in 1616 patients (90.0 %)
- 777 (43.3 %) had a liver biopsy.

BSG Guidelines :

- MRCP should be the principal imaging modality for the investigation of suspected PSC. ERCP should be reserved for patients with biliary strictures
- Liver biopsy reserved for SD PSC, overlap or unclear diagnosis

Liver Disease Risk stratification

- 785 patients (43.7 %) had not undergone disease staging or risk stratification within the last 2 years
- where performed, it was most commonly by transient elastography (n = 645, 78.7 %).

BSG Guidelines :

- Liver disease risk stratification is recommended based on non-invasive assessment.

Fatourou et.al. Gut 2021;**70:**A9

AASLD Guidelines

LS measurement by TE or MRE is the preferred method and should be done at diagnosis and regularly during follow-up

- **EASL Guidelines**

  Liver elastography and/or serum fibrosis tests at least 2 to 3 years are recommended (Strong recommendation)

AASLD PSC Guidelines Hepatology 2022
EASL PSC guidelines J Hep 2022

Ursodeoxycholic acid (UDCA)

- 931 patients (51.9 %) received non-licensed therapy with UDCA
- UDCA dose (n=745)

➢ <10mg/kg, n=234 (31.4%)

➢ 10-15mg/kg, n=367 (49.3%)

➢ >15mg/kg, n=144 (19.3%)

BSG Guidelines :UDCA should not used for the routine treatment of newly diagnosed PSC or for colorectal cancer/cholangiocarcinoma prevention

EASL guidelines : UDCA 15-20 mg per kg daily (weak recommendation)

AASLD guidelines : patients with persistently elevated ALP/gGT can be considered for UDCA 13-23 mg per kg daily and treatment should be continued if there is reduction or normalisation of ALP or improvement of symptoms (weak recommendation)

AASLD PSC Guidelines Hepatology 2022
EASL PSC guidelines J Hep 2022

Surveillance for biliary tract Ca

- Surveillance for biliary tract cancer was not undertaken in 515 patients (28.7 %)
- When performed, most commonly by ultrasound (US) (n = 568, 47.1 %)
- or alternating MRCP/US (n = 429, n = 35.6 %).
- Ca 19 - 9 was utilised in 730 patients (40.6%)

BSG Guidelines :

- annual ultrasound scan of the gallbladder should be performed
- Routine measurement of serum CA19.9 is not recommended for surveillance for cholangiocarcinoma

Surveillance for cholangiocarcinoma

- retrospective data from 2975 PSC patients from 27 European Centres
- Most centres used ultrasound (US) and/or magnetic resonance imaging (MRI). Two centres used scheduled endoscopic retrograde cholangiopancreatography (ERCP) in addition to imaging for surveillance purposes
- Scheduled imaging leads to improved survival

Berguist et.al. Liver International 2023

- Retrospective data, Mayo clinic (1995 to 2015)
- a total of 79 of 830 PSC patients were diagnosed with HBCa.
- Patients in the surveillance group had significantly higher 5-year overall survival (68% versus 20%, respectively; $P < 0.001$)

Ali et al. Hepatology 2018

AASLD guidelines :

- CCA and gallbladder Ca surveillance should be performed annually including preferably MRI/MRCP with or without Ca 19-9
- Patient with cirrhosis should have 6 monthly HCC surveillance
- Cholecystectomy for polyps >8mm

EASL guidelines :

- Surveillance with US and/or MRCP for CCA and gallbladder malignancy at least yearly in patients with large duct PSC
- In patients with cirrhosis 6 monthly surveillance is advised
- Cholecystectomy for polyps >8mm
- Ca 19-9 is not suggested for surveillance

UK-PSC

Inflammatory bowel disease Demographics & Management

IBD Demographics

UK-PSC

- Concurrent IBD was present in 1264 patients (70.4 %)
- 256 (20.3 %) had a colectomy.
- Where classified, pancolitis (Montreal classification E3) was the commonest disease distribution (673 / 939, 71.7 %)
- 1.6 % (n = 15) having isolated ileal disease.

Fatourou et.al. Gut 2021;**70:**A9

IBD management

- Most patients with IBD were followed up by an IBD specialist n=616 (48.7%)
- 266 (21.1%) were followed by a general gastroenterologist
- 236 (18.7%) by a hepatologist
- 15 (1.2%) patients were followed in a joint IBD/Hepatology clinic.

Fatourou et.al. Gut 2021;**70:**A9

Colonoscopy surveillance

- Of those <u>without</u> documented IBD diagnosis, only 303/507 (59.7%) had this excluded by colonoscopy and biopsies.
- Among those with colitis without previous colectomy (n=743),

580 (78.1%) underwent annual colonoscopic surveillance.

BSG Guidelines:

- Colitis should be sought in all patients with PSC using colonoscopy and colonic biopsies
- In the presense of IBD, annual colonoscopic surveillance

Colonoscopy surveillance method

- Among those with colitis without previous colectomy,

who underwent annual colonoscopic surveillance n=580 (78.1%)

✓30 (5.2%) had dye spray,

✓230 (39.7%) had biopsies and dye spray,

✓252 (43.4%) had protocol biopsies alone.

BSG Guidelines:
Pancolonic dye spraying with targeted biopsy of abnormal areas is recommended.

EASL guidelines :

- Ileo-colonoscopy with biopsies at time of diagnosis regardless of presence of inflammation/lesions
- Annual colonoscopy surveillance with biopsies in IBD PSC
- ESGE: Targeted biopsies with dye-spray in IBD-PSC as a standard surveillance investigation

AASLD guidelines :

High definition or dye spray colonoscopy with biopsies yearly or every 2 years

AASLD PSC Guidelines Hepatology 2022
EASL PSC guidelines J Hep 2022

Colonoscopy surveillance

- No Difference between IBD or joint care vs hepatologist/general gastroenterologists
- Age <40 was associated with poorer compliance to annual colonoscopy surveillance, ($p=0.023$)

Conclusions

UK-PSC

Unwarranted variation in the care of PSC patients across the UK

- Risk stratification for liver disease
- Surveillance for biliary tract Cancer / gallbladder Cancer
- Exclusion of colitis at the time of PSC diagnosis
- Colonic cancer surveillance in patients with IBD-PSC

Fatourou et.al. Gut 2021;**70:**A9

Conclusions/Recommendations

UK-PSC

- Lack of uniformity highlights the need for better education of clinicians about PSC management and radiological diagnosis
- EASL guidelines recommend expert opinion at time of diagnosis and referral to experienced centre with MDT input (imaging review and participation in clinical trials)
- Potential role of clinical networks for rare liver diseases within the UK.

EASL PSC guidelines J Hep 2022

Conclusions

- Conflicting areas between EASL, AASLD and BSG guidelines as far as CCA surveillance and UDCA treatment
- BSG guidelines should be revised, as far as surveillance of CCA is concerned, and re-audited
- PSC support patient survey (n=190) found that 84%–90% considered it 'extremely important' to improve cancer screening for people with PSC.

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