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October 03, 2024 Protect yourself this fall with essential vaccines for flu, COVID-19 and RSV. Learn about timing, tips, and recommendations for a healthier season. 0 ratings0% found this document useful (0 votes)10 viewsSaveSave 22a AASLD Practice Guidance on Primary Sclerosing ... For Later0%0% found this document useful, undefined Share — copy and redistribute the material in any medium or format for any purpose, even commercially. Adapt — remix, transform, and build upon the material for any purpose, even commercially. The licensor cannot revoke these freedoms as long as you follow the license terms. Attribution — You must give appropriate credit , provide a link to the license, and indicate if changes were made . You may do so in any reasonable manner, but not in any way that suggests the licensor endorses you or your use. ShareAlike — If you remix, transform, or build upon the material, you must distribute your contributions under the same license as the original. No additional restrictions — You may not apply legal terms or technological measures that legally restrict others from doing anything the license permits. You do not have to comply with the license for elements of the material in the public domain or where your use is permitted by an applicable exception or limitation . No warranties are given. The license may not give you all of the permissions necessary for your intended use. For example, other rights such as publicity, privacy, or moral rights may limit how you use the material. Acute Liver Failure, Management Acute-on-Chronic Liver Failure and the Management Alcohol-Associated Liver Disease Ascites, Spontaneous Bacterial Peritonitis and Hepatorenal Syndrome, Management Autoimmune Hepatitis, Management Drug, Herbal, and Dietary Supplement-induced Liver Injury Hemochromatosis, Management Hepatic Encephalopathy Hepatitis B, Chronic Hepatitis C, Guidance Hepatocellular Carcinoma, Management Liver Biopsy Liver Transplantation, Evaluation of the Adult Patient Liver Transplantation, Evaluation of the Pediatric Patient Long-Term Management of the Adult Liver Transplant AASLD strives to review and update its practice guidelines every five (5) years, as necessary. Users are cautioned that in the interim, scientific and medical developments may supersede or invalidate, in whole or in part, specific recommendations in any guideline. A guideline is considered to be "inactive" if it has not been updated by AASLD in at least five (5) years, and for this reason particular care must be exercised in placing reliance on an inactive guideline. AASLD commissions and provides financial support for the formulation and production of practice guidelines/guidances by volunteer experts. Financial support from commercial entities or the pharmaceutical industry is not accepted for the development of AASLD practice guidelines. The 2023 practice guidance on primary sclerosing cholangitis (PSC) and cholangiocarcinoma (CCA) of the American Association for the Study of Liver Diseases (AASLD) came as a needful update to the previous 2010 guidelines on PSC, with a first-time inclusion of dedicated guidance on the diagnosis and management of CCA (1,2). This data-supported approach developed by consensus of an expert panel, provides guidance statements based on analytical review of the relevant literature. The production of guidance rather than guidelines relates to the paucity of sufficient randomized controlled trials suitable for systematic review and meta-analysis (1). The publication is comprehensive, illustrative of remarkable progress since the 2010 guidelines, and will be a useful tool in providing high-quality care for these patient groups. Aside from the significant amount of new information and helpful recommendations, there is room to identify current gaps and future directions. The new terminology and clarifications about biliary strictures in PSC, including dominant, high-grade and relevant stricture, are valuable as they cover pre-existing gaps while promoting the use of a common language worldwide, as recently recommended by the International PSC Study Group (1,3). High-quality MRCP is recommended as the gold standard for diagnosis, while avoiding ERCP for diagnostic purposes is highlighted, owing to the increased risk of complications. The recently published guidelines of the European Association for the Study of the Liver (EASL) are less stringent on this matter but in agreement that MRCP is the preferred diagnostic tool (4). While liver biopsy is fundamental to confirm a diagnosis of small-duct PSC or autoimmune hepatitis overlap (3), it is not recommended for fibrosis staging in clinical practice (1,4). However, the role of liver histology in PSC has been recently revisited and is now considered essential in the regulatory approval process for new therapeutic strategies (5). In the current landscape of intense development of surrogates of clinical endpoints, it is worth mentioning the recent data on collagen proportionate area (CPA), a quantitative method to measure the proportion of fibrosis on liver biopsy samples using digital image analysis. Further to the traditional histology scores, CPA provides useful information for risk-stratification and prognostication in PSC, and may be particularly useful in the clinical trials setting, for assessment of response-to-treatment and regression of fibrosis (6). The utility of liver stiffness assessment by transient elastography or magnetic resonance elastography to estimate liver fibrosis and predict clinical outcomes, is increasingly recognised (4,7). Albeit not discussed in the new guidance, increasing evidence shows that the more recently developed elastographic techniques such as point- and 2D-/3D-shear wave elastography can be reliable for assessment of liver fibrosis and risk-stratification in PSC. Recent data on spleen stiffness as a predictor of presence of oesophageal varices in PSC are also encouraging (8). Clinical prediction scores designed for PSC do not have generalised reliability and applicability; the 2023 practice guidance provides a useful diagram to guide the clinician in choosing, based on individual features (1). Recommendations on the use of ursodeoxycholic acid (UDCA) in PSC have been inconsistent over time, and its potential to improve clinical outcomes remains unclear. However, a moderate dose of UDCA is now suggested as a possible treatment for patients not interested in or ineligible for clinical trials, since it may improve serum liver tests and surrogate markers of prognosis. The suggested dose, however, differs between the AASLD guidance (13–23 mg/kg/day) and the recent EASL guidelines (15–20 mg/kg/day) (1,4). As controversy still exists on the topic, further data is required to establish optimal doses. The 2023 AASLD guidance enumerates various pharmacological treatments currently investigated in clinical trials. It is worth also noting that faecal microbiota transplant has shown good safety profile and some preliminary efficacy signals in a recent pilot study, and is currently being further investigated (9). The new guidance is the first ever to recommend the onset of colorectal cancer surveillance in patients with inflammatory bowel disease at the age of 15 years (1). Notably, it recommends considering cholecystectomy for gallbladder polyps >8 mm, in contrast with the European guidelines from 2017, which suggested a cut-off size of >6 mm (10), while the EASL guidelines recommend it for polyps ≥8 mm (4). Furthermore, EASL suggests assessment of smaller polyps with contrast-enhanced ultrasound, and consideration of cholecystectomy if they enhance (4). The new guidance appropriately differentiates de novo CCA and PSC-associated CCA, which owing to distinctive features should be seen indeed as different entities (11). The provided diagnostic algorithm for relevant strictures in PSC is highly useful (1). Some aspects are particularly interesting, including initial suspicious cytology warranting ERCP in 3 months, regardless of fluorescent in situ hybridization (FISH) results (1). It would be useful to assess the proportion of PSC patients ultimately diagnosed with CCA after recommended interval ERCP at 3 months and, subsequently, the proportion of those eligible for curative-intent treatment. Furthermore, clear recommendations are warranted for when initial negative/suspicious cytology with FISH polysomy is followed by ERCP revealing suspicious cytology with either FISH polysomy or negativity. Importantly, AASLD recommends that FISH should be obtained in all patients with suspected perihilar CCA (pCCA) or distal CCA (dCCA) (1). While FISH has undoubtedly increased the diagnostic accuracy of biliary brushings cytology, it is not uniformly available for this purpose, including in prominent centres. It has to be underlined that the ongoing progress with liquid biopsies in different fronts, including diagnosis (12), might result in the inclusion of this technique in future guidance and at various stages within the diagnostic algorithms. A few remarks on the valuable therapeutic algorithms for the three topographic categories of CCA can be drawn. A guidance statement explains that for unresectable liver-limited intrahepatic CCA (iCCA) orthotopic liver transplantation (OLT) should be considered under research protocols only (1), while the schematic therapeutic algorithm recommends referral to a liver transplant centre for unresectable iCCA ≤2 cm. Although iCCA is still largely considered a contraindication for OLT, recent promising outcomes have led to the consensus statement recommendation by the European Network for the study of CCA that OLT should be considered as a potentially curative option especially in patients with very early stage unresectable iCCA (≤2 cm) and cirrhosis (13). The guidance recommendation for resectable pCCA is “surgery followed by adjuvant chemotherapy”. Notably, in PSC-associated pCCA, surgical resection is most frequently precluded owing to various parameters including a higher rate of multifocal CCA, skip cancer lesions, poor quality of the liver parenchyma and hepatic dysfunction with reduced regenerative capacity (11,13). In contrast, the option of neoadjuvant chemoradiotherapy followed by OLT is increasingly being recognised as the optimal treatment strategy and remains among the inclusion criteria of the Mayo Clinic protocol. The outcomes are superior compared to de novo pCCA (11,13), likely due to earlier detection in PSC owing to routine surveillance (1) and a possible higher responsiveness to radiation therapy on a background of PSC (11,13). Extension into the distal bile duct is an established reason for consideration of combined OLT and pancreatoduodenectomy (11,13). The emphasis of the AASLD group on the Mayo Clinic protocolised approach to early pCCA is important, particularly as OLT for pCCA remains contraindicated in many countries despite the evidence of very good outcomes, on the background of the relative scarcity of donor organs. The TRANSPHIL trial (NCT02232932), a French prospective open-label randomised multicentre comparative study ongoing since 2012, is aiming to clarify whether the best treatment for resectable pCCA is neoadjuvant chemoradiotherapy followed by OLT or standard of care liver and bile duct resection. Should previous positive outcomes in favour of OLT be confirmed, this will lead to a true paradigm shift in the management of pCCA (11). Notably, the trial lists PSC among the exclusion criteria for enrolment. Even without a statement regarding the role of lymphadenectomy, the practice guidance discusses that for iCCA in general, surgery involves liver resection and portal lymphadenectomy (1). This approach is not uniformly accepted and the role of lymphadenectomy in this setting as a standard part of the surgical resection is under ongoing debate. Our recent meta-analysis showed that lymphadenectomy did not improve overall and disease-free survival, except in Japanese studies. However, lymph node metastases were found in 27.7% of patients undergoing lymphadenectomy, suggesting that it may aid in adequate staging (14). Statements regarding the adequacy of lymphadenectomy in pCCA and dCCA are lacking. There is controversy on the matter in pCCA, with no established minimal number of harvested lymph nodes for histopathological assessment, while for dCCA, a minimum of 12 lymph nodes has been suggested as appropriate for accurate staging (15). Firmer recommendations are anticipated in the future on this topic. When CCA extends both distally and towards the liver, attempts at hepatopancreatoduodenectomy have been challenging with high perioperative morbidity and mortality (97.4% and 26%, respectively, in recent series) and 5-year overall survival of 17.9–49.2% (13). Accordingly, this procedure may be suitable to consider as a potentially curative approach only in very carefully selected fit patients. As underlined by the 2023 guidance, laparoscopic liver surgery is a safe approach for liver cancer (1). Notably, the rapid accumulation of evidence with robotic surgery may soon define its place in the treatment of iCCA. In summary, the new AASLD practice guidance on PSC and CCA came at a suitable time and, compared with the guidelines from 2010, is highly indicative of the remarkable progresses made in understanding these diseases and their management. While it is expected to function as a valuable tool for specialists in the relevant fields, the current pace of progress is promising that further important outstanding questions may be answered in the near future. The article’s supplementary files as Funding: None.Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. 1.Bowlus CL, Arrivé L, Bergquist A, et al. AASLD practice guidance on primary sclerosing cholangitis and cholangiocarcinoma. Hepatology 2023;77:659-702. 10.1002/hep.32771 [DOI] [PubMed] [Google Scholar] 2.Chapman R, Fevery J, Kalloo A, et al. 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[Google Scholar] This section collects any data citations, data availability statements, or supplementary materials included in this article. The article’s supplementary files as Iowa, Minnesota, Wisconsin AASLD develops evidence-based practice guidelines and practice guidances which are updated regularly by a multi-disciplinary panel of experts, including hepatologists, and include recommendations of preferred approaches to the diagnostic, therapeutic, and preventive aspects of care. Guidelines for the management of primary sclerosing cholangitis (PSC) have recently been published by both the European Association for the Study of the Liver (EASL) and the American Association for the Study of Liver Diseases (AASLD). The current review focuses on the management of PSC, based on these guidelines. There is no established medical therapy for PSC. The role for UDCA in slowing the disease progression and improving survival is as yet unclear, and there are no specific recommendations for the general use of UDCA in this condition. Guidelines recommend that dominant bile duct strictures with significant cholestasis should be treated with biliary dilatation, with or without stenting. Prospective studies to define type, duration, optimal frequency and long-term effects of endoscopic therapy are needed. Liver transplantation is recommended for end stage disease and has excellent results. PSC patients with dysplasia in biliary brush cytology specimens should also be considered for transplantation. There is no evidence-based algorithm for the follow-up of PSC patients, but some regular investigations are recommended (surveillance colonoscopies in patients with IBD and ultrasound to detect gallbladder mass lesions). If the ducts that lead from the liver to the duodenum sustain enough damage, or experience repeated bouts of cholangitis, gallstones, or other conditions that affect this tissue, the defense mechanism of the body is to develop scar tissue, which can then hinder the movement of bile secretions. Increased scarring leads to more damage, and forces bile back into the liver, which damages liver cells. What Causes PSC? While many conditions are implicated in the development of PSC, no root cause has yet been determined. It is thought that genetics may play a role in PSC, as well as possibly viruses and bacteria. Additionally, it has been noted that many patients with PSC also have immune problems and/or suffer from ulcerative colitis. What are the symptoms of PSC? Primary sclerosing cholangitis develops over time, as repeated attacks of gallstones and cholangitis cause the hardening of the duct walls. Although it can affect people of all ages, it primarily affects men over the age of forty. Symptoms may include: Fatique Jaundice in the skin or eyes Chills and fever Diagnosis Blood tests to check levels of liver enzymes are the first step in diagnosing primary sclerosing cholangitis. Doctors confirm the diagnosis using cholangiography, which provides pictures of the bile ducts. Cholangiography can be performed in the following ways: ERCP — an endoscope (a long, flexible, lighted tube) enters the mouth, slides down the esophagusm, traverses through the stomach, and finally into the duodenum (first part of the small intestine) where dye can be injected (into the bile ducts) and X-rays are taken. ERCP also can be used to take a tissue sample or to treat blocked ducts. Percutaneous transhepatic cholangiography — a needle is inserted through the skin and a thin tube is placed into a duct in the liver. Dye is injected through the tube and x-rays are taken. MRCP/MRI — uses radio waves and magnets to scan internal organs and tissues, including the bile ducts without the use of x-rays or the insertion of instruments into the body. Other testing may include ultrasound exams and a liver biopsy. Ultrasound uses sound waves to create images of organs inside the body. A biopsy involves removal of a small piece of tissue for examination with a microscope. Treatment Although researchers have studied many treatments, none has been shown to cure or slow the progress of primary sclerosing cholangitis. Treatment of PSC aims to relieve symptoms and manage complications. Medical treatment may include various medications to relieve itching, antibiotics to treat infections, and vitamin supplements. Instruments passed through an endoscope during ERCP can help open blocked bile ducts. Liver transplantation may be an option if the liver begins to fail. Complications Primary sclerosing cholangitis can lead to various complications, including: deficiencies of vitamins A, D, E, and K infections of the bile ducts cirrhosis (an extensive scarring of the liver) liver failure bile duct cancer Summary Primary sclerosing cholangitis inflames, scars, and blocks bile ducts inside and outside the liver. When these bile ducts become blocked, bile builds up in the liver and causes damage to the liver cells. PSC can lead to vitamin deficiencies, infections, bile duct cancer, cirrhosis, liver failure, and the possible need for a liver transplant. The exact cause of PSC is not known, but many people with PSC also have ulcerative colitis, an inflammatory bowel disease. Medications can be used to treat symptoms and complications of PSC. In patients with suspected PSC, a 3D MRI/MRCP with T1w and T2w axial images and contrast enhancement should be obtained to evaluate for cholangiographic features of PSC, including intrahepatic and/orextrahepatic strictures alternating with normal or slightly dilated segments. In patients with suspected PSC and a normal, high-quality MRI/MRCP, liver biopsy should be considered to rule out small-duct PSC. Patients with an equivocal MRI/MRCP should be referred to an experienced center for consideration of a repeat high-quality MRI/MRCP or liver biopsy. A repeat MRI/MRCP may be considered in 1 year if the diagnosis remains unclear. ERCP should be avoided for the diagnosis of PSC. In all patients with possible PSC, serum IgG4 levels should be measured to exclude IgG4-sclerosing cholangitis. A liver biopsy should not be performed in patients with typical cholangiographic findings on MRI/MRCP, except when there is concern for autoimmune hepatitis overlap. Ileocolonoscopy with biopsies should be performed in patients with a new diagnosis of PSC and no previous diagnosis of IBD. In patients without IBD, subsequent ileocolonoscopy should be considered at 5-year intervals or whenever symptoms suggestive of IBD occur. Patients with small-duct PSC should be monitored by MRI/MRCP every 3–5 years for the development of large-duct disease. Risk stratification and fibrosis staging should be done at diagnosis of PSC and regularly during follow-up. Clinical risk tools can be considered for this purpose, but specific probabilities of events should be interpreted with caution in the individual patient. Liver stiffness measurement by transient elastography or magnetic resonance elastographyis currently the preferred method for estimation of fibrosis stage in PSC. Liver biopsy is not recommended for fibrosis staging in clinical practice.