PERSONAL

EXPERIENCE

EXPECTATIONS

KNOWLEDGE

PEEK

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PRIMARY BILIARY CHOLANGITIS

AUSTRALIA 2025

Clinical Pathway REVIEW

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METHODOLOGY

- **Review of published clinical guideline recommendations at different time points to understand progress in management**
  - Literature scan of clinical guideline recommendations published in medical journals before 2020
  - Literature scan of clinical guideline recommendations published in medical journals after 2020
- **Review of PEEK study results in the context of clinical guideline recommendations**
  - Data extracted across the domains of diagnosis, treatment, psychosocial support, lifestyle modifications and long-term follow-up

Published clinical guideline recommendations before 2020

History & Symptoms

Early clinical guidance, such as from the European Association for the Study of the Liver (EASL, 2017), emphasizes that PBC should be suspected in patients presenting with persistent cholestatic serum abnormalities—notably elevated alkaline phosphatase—and symptoms like pruritus or fatigue even when initial nonspecific (EASL, 2017). The 2018 AASLD Practice Guidance similarly underscores the importance of biochemical cholestasis (e.g., elevated ALP), in conjunction with serologic markers, for recognizing PBC (Lindor et al., 2018).

Differential Diagnosis

The 2018 AASLD guidance specifies that a diagnosis requires at least two of the following: cholestatic biochemistry, antimitochondrial antibody (AMA) positivity, or characteristic histopathology—thus clearly delineating differential conditions such as cholestatic drug reactions, biliary obstruction, sarcoidosis, autoimmune hepatitis, and primary sclerosing cholangitis (Lindor et al., 2018). These criteria help clinicians systematically exclude overlapping or mimic conditions in the diagnostic workflow.

Diagnostic Tests

According to EASL (2017), after initial suspicion based on symptoms and cholestatic liver tests, further diagnostic evaluation should include imaging (e.g., MRCP) to exclude obstructive causes, with liver biopsy reserved for cases with AMA negativity or suspected overlap syndromes—providing guidance on when invasive testing is warranted (EASL, 2017). Similarly, the AASLD guidance (Lindor et al., 2018) outlines the triad for diagnosis, advising histopathology when serologic and biochemical criteria are inconclusive.

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Published clinical guideline recommendations after 2020

History & Symptoms

The 2021 AASLD practice guidance update underscores that many cases of PBC are diagnosed incidentally, often in asymptomatic individuals referred for routine testing, although fatigue and pruritus remain the most frequently reported symptoms (Lindor et al., 2022). This reflects an evolution in clinical presentation awareness, recognizing that disease onset may be silent, with symptoms emerging as the disease progresses.

Differential Diagnosis

The updated guidance reinforces that diagnosis hinges on at least two of the following: cholestatic serum markers, antimitochondrial antibodies (AMA), and histologic evidence (Lindor et al., 2022). This triad continues to assist clinicians in confidently distinguishing PBC from other conditions—such as primary sclerosing cholangitis, autoimmune hepatitis, and cholestatic drug reactions—ensuring accuracy in differential diagnosis.

Diagnostic Tests

More recently, the 2021 Chinese Society of Hepatology guidelines provide a comprehensive diagnostic algorithm that integrates clinical presentation, laboratory evaluations (liver biochemistries, AMA serology), imaging studies, and histology (You et al., 2023). This multifaceted approach ensures that clinicians can effectively confirm PBC, monitor disease progression, and tailor diagnostic strategies to regional practices and patient demographics.

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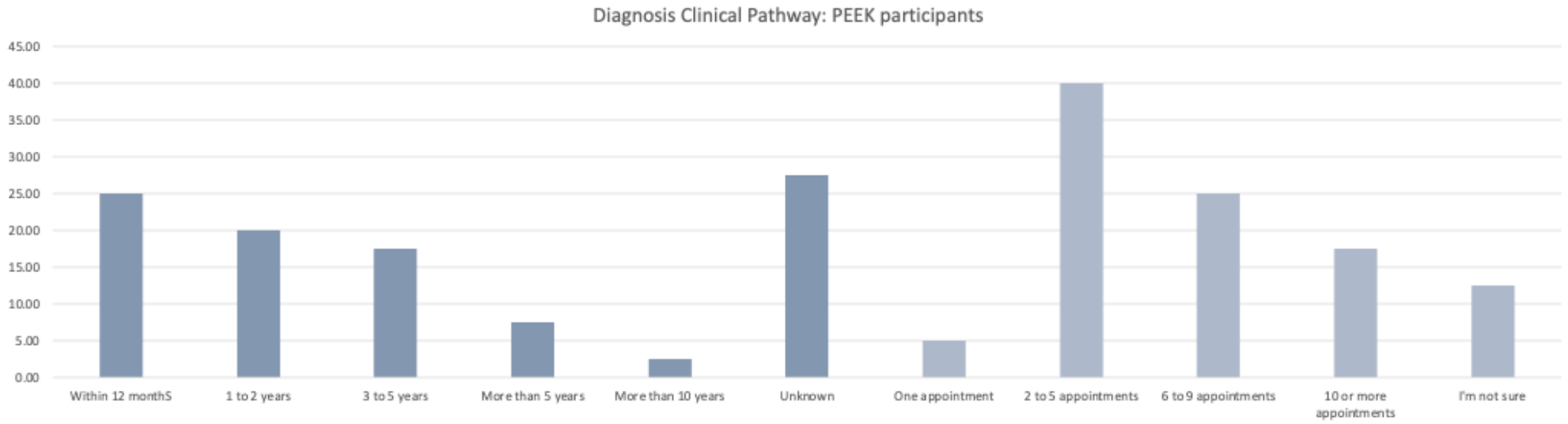
Published clinical guideline recommendations

Domain	Before 2020 (EASL 2017, AASLD 2018)	After 2020 (AASLD 2022, CSH 2021)
History & Symptoms	Emphasis on *symptomatic presentation* (pruritus, fatigue) or detection during abnormal liver biochemistry; symptoms often trigger testing	Greater acknowledgment of *asymptomatic presentations* found during routine labs; symptoms may develop later (Lindor et al., 2022)
Differential Diagnosis	Diagnosis requires ≥2 of 3: cholestatic liver biochemistry, AMA positivity, characteristic histology; clear exclusion of PSC, AIH, drug injury	Same ≥2 of 3 core criteria, but stronger emphasis on structured *diagnostic algorithms* and exclusion pathways (You et al., 2023)
Diagnostic Tests	AMA testing as primary serologic marker; imaging (e.g., MRCP) only to rule out obstruction; biopsy reserved for AMA-negative or overlap cases	Maintains AMA as primary marker, but integrates broader use of imaging + non-invasive fibrosis tools; biopsy role unchanged but algorithms more explicit

DIAGNOSIS

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DIAGNOSIS

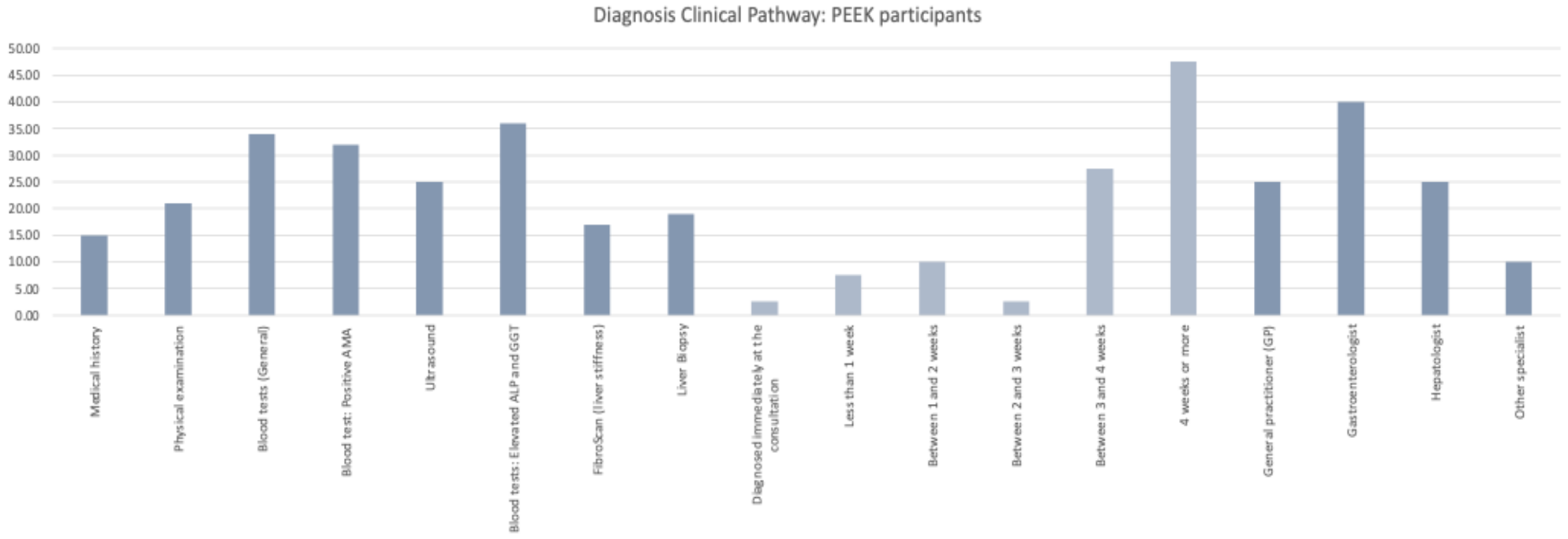


Time from symptoms to diagnosis

Number of appointments to confirm diagnosis

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DIAGNOSIS

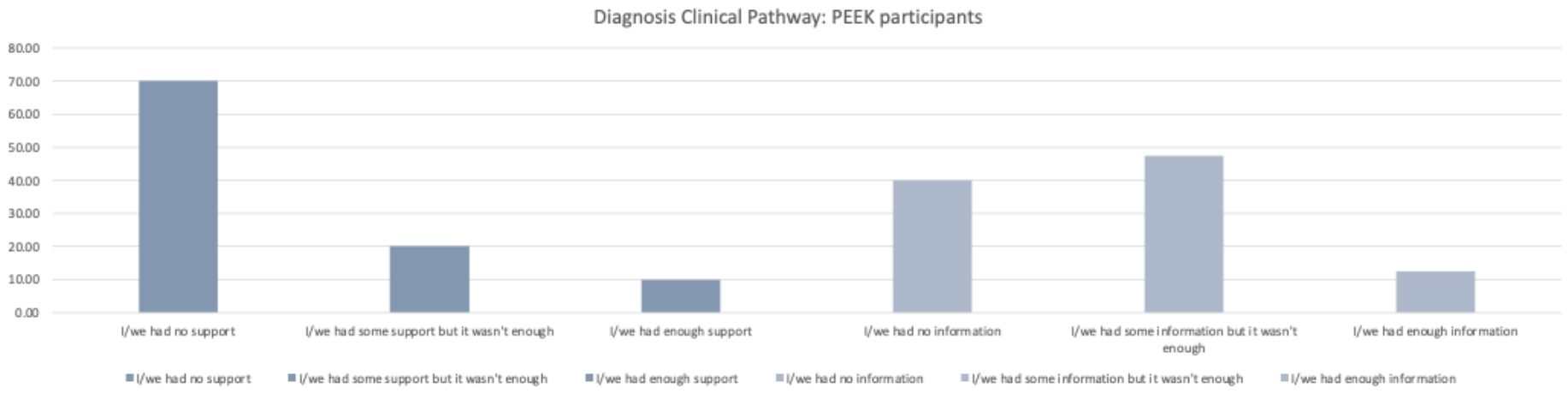


Diagnostic tests

Time from tests to diagnosis

Diagnosis delivered by

DIAGNOSIS



Support at diagnosis

Information at diagnosis

Published clinical guideline recommendations before 2020

Treatment options

First-line treatments for PBC was ursodeoxycholic acid (UDCA) at a dose of 13–15 mg/kg/day, continued indefinitely, as recommended by both the European Association for the Study of the Liver (EASL, 2017) and the American Association for the Study of Liver Diseases (AASLD, 2018). UDCA improves biochemical markers, delays histological progression, and increases transplant-free survival (EASL, 2017; Lindor et al., 2018). For patients with an inadequate biochemical response after 12 months on UDCA, second-line options were advised.

In the U.S. and Europe, this included obeticholic acid (OCA)—approved for use either in combination with UDCA or as monotherapy for UDCA-intolerant patients—based on its effect on reducing alkaline phosphatase and bilirubin levels (Lindor et al., 2018). The EASL guidelines also acknowledged off-label use of fibrates (bezafibrate, fenofibrate) as adjunctive therapy in partial responders, supported by small-scale trials showing improvement in biochemical response, though not yet approved for this indication (EASL, 2017). Supportive therapy recommendations included cholestyramine or rifampin for pruritus, fat-soluble vitamin supplementation for malabsorption, and bone protection for osteoporosis risk (EASL, 2017). Liver transplantation was advised for patients with end-stage liver disease, intractable pruritus, or severe fatigue unresponsive to medical therapy (EASL, 2017; Lindor et al., 2018).

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Published clinical guideline recommendations after 2020

Treatment options

Current guidance maintains ursodeoxycholic acid (UDCA) 13–15 mg/kg/day as first-line therapy for all patients and recommends assessing biochemical response at ~12 months to decide on escalation (Lindor et al., 2022; You et al., 2023). For UDCA non-responders or UDCA-intolerant patients, obeticholic acid (OCA) is recommended as second-line therapy, avoiding use in advanced/decompensated cirrhosis in light of safety restrictions; initiation typically starts at 5 mg/day with careful monitoring (Lindor et al., 2022).

Multiple post-2020 guidelines also support considering fibrates (especially bezafibrate; fenofibrate where available) as off-label adjuncts to UDCA in partial responders, with caution in decompensated disease and attention to renal function and drug interactions (APASL/You et al., 2022; You et al., 2023). Symptom-directed care remains integral: bile-acid sequestrants (e.g., cholestyramine) first-line for pruritus, with rifampin, opioid antagonists, or SSRIs as subsequent options; ensure bone health measures and fat-soluble vitamin support when indicated (You et al., 2023). Liver transplantation continues as definitive therapy for end-stage disease or refractory, life-limiting symptoms (APASL/You et al., 2022; You et al., 2023).

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Published clinical guideline recommendations

Therapy Area	Before 2020 *(EASL 2017; AASLD 2018)*	After 2020 *(AASLD 2022; APASL 2022; CSH 2021)*
First-line therapy	UDCA 13–15 mg/kg/day lifelong for all patients; assess biochemical response after 12 months.	Same UDCA recommendation and dose; emphasis on standardized response criteria and routine 12-month reassessment.
Second-line for inadequate UDCA response	OCA approved for combination therapy or monotherapy in UDCA-intolerant patients; dosing typically starts at 5 mg/day; limited safety warnings.	OCA recommended only if no decompensated cirrhosis (Child-Pugh B or C) due to FDA safety alerts; start 5 mg/day, monitor closely; dose adjustment for hepatic impairment.
Alternative/adjunctive therapy	Off-label fibrates (bezafibrate, fenofibrate) suggested for partial UDCA responders - limited evidence, not guideline standard.	Stronger support for bezafibrate as adjunct in partial responders (off-label in many regions), caution in advanced disease; fenofibrate mentioned where available.
Pruritus management	Stepwise: cholestyramine - rifampin - naltrexone or SSRIs if refractory; supportive measures advised.	Same sequence, but more explicit on early escalation for refractory pruritus and multidisciplinary symptom care.

TREATMENT

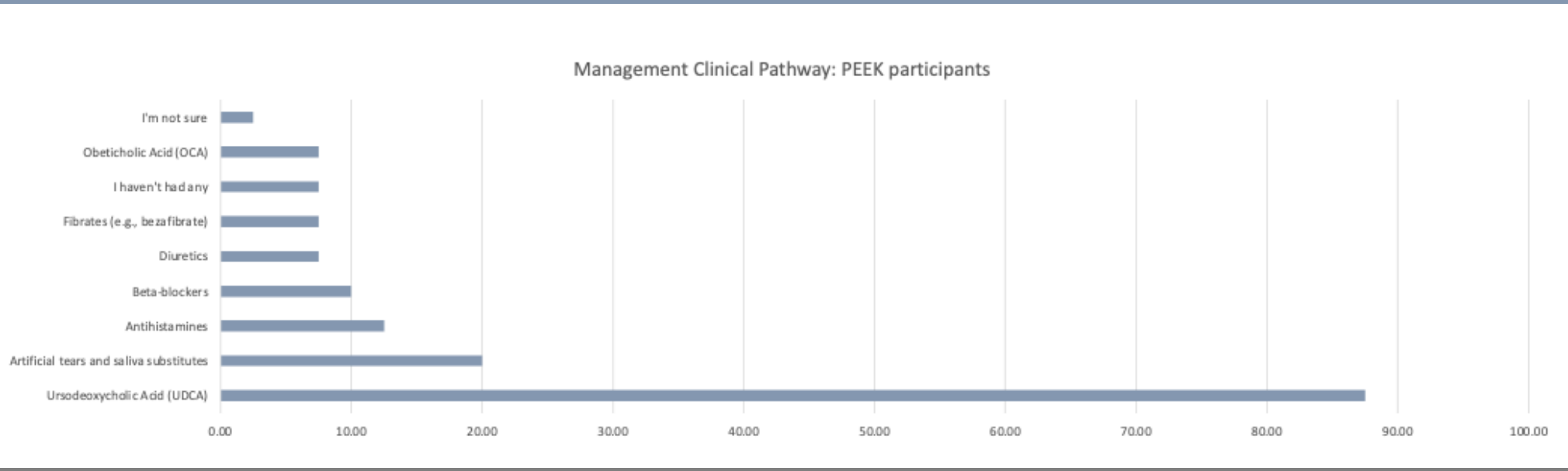
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Published clinical guideline recommendations

Therapy Area	Before 2020 *(EASL 2017; AASLD 2018)*	After 2020 *(AASLD 2022; APASL 2022; CSH 2021)*
Supportive care	Bone health, fat-soluble vitamin supplementation, lipid monitoring, osteoporosis prevention.	Same, with added emphasis on fracture risk assessment and periodic re-evaluation of nutritional status.
Liver transplantation	Indicated for decompensated cirrhosis, severe pruritus, or fatigue unresponsive to therapy.	Same, but newer guidance highlights transplant for refractory pruritus even in compensated disease when quality of life is severely impaired.

TREATMENT

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Treatments accessed

TREATMENT

Published clinical guideline recommendations before 2020

Psychological Support

Before 2020, both the European Association for the Study of the Liver (EASL, 2017) and the American Association for the Study of Liver Diseases (AASLD, 2018) recognized that PBC symptoms such as chronic fatigue, pruritus, and disease progression could significantly affect mental health and quality of life. While these guidelines did not prescribe a specific psychological therapy, they emphasized the importance of acknowledging the psychosocial impact of PBC and ensuring multidisciplinary care, which could include counseling, mental health referral, and patient support groups (EASL, 2017; Lindor et al., 2018). They also noted the need for open communication between clinicians and patients regarding symptom burden, and recommended proactive screening for depression and anxiety given their higher prevalence in PBC populations.

Lifestyle Modifications

Guidelines before 2020 recommended general liver health measures to optimize wellbeing and slow disease progression risk. This included abstinence from alcohol, maintaining a balanced diet rich in nutrients (with particular attention to fat-soluble vitamin intake in advanced cholestasis), and regular weight-bearing exercise to reduce osteoporosis risk (EASL, 2017). Smoking cessation was encouraged both for general health and possible reduction in autoimmune disease triggers. Sunlight exposure and vitamin D supplementation were recommended in patients at risk for deficiency. Additionally, patients were advised to maintain adequate hydration, limit hepatotoxic medications, and follow vaccination schedules for hepatitis A and B if not immune (EASL, 2017; Lindor et al., 2018).

SUPPORTIVE CARE & EDUCATION

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Published clinical guideline recommendations after 2020

Psychological Support

Recent guidance emphasizes structured, patient-centred symptom care for pruritus, fatigue, sleep disturbance, mood symptoms, and health-related quality of life, recommending routine screening for anxiety/depression and sleep problems, clear education about symptom mechanisms, and timely referral to mental-health and supportive services (e.g., counseling, CBT-based approaches) when symptoms impair daily functioning (Lindor et al., 2022; You et al., 2022; You et al., 2023). Clinicians are encouraged to validate symptom burden, set shared goals, and consider multidisciplinary management (hepatology, dermatology, psychology, specialist nursing) as part of ongoing care, alongside escalation of pruritus and fatigue therapies when needed (Lindor et al., 2022; You et al., 2023)

Lifestyle Modifications

Post-2020 guidelines continue to recommend alcohol avoidance, smoking cessation, and a balanced diet with attention to vitamin D and calcium for bone health, plus weight-bearing exercise tailored to fatigue levels; they also advise vaccination against hepatitis A/B when non-immune and review of potentially hepatotoxic or pruritus-worsening medications (Lindor et al., 2022; You et al., 2023). Practical measures for itch (e.g., emollients, avoiding heat/hot baths, loose clothing), sleep hygiene, gradual activity pacing, and periodic reassessment of nutritional status and fracture risk are highlighted to complement pharmacologic therapy and long-term UDCA-based management (You et al., 2022; You et al., 2023).

SUPPORTIVE CARE & EDUCATION

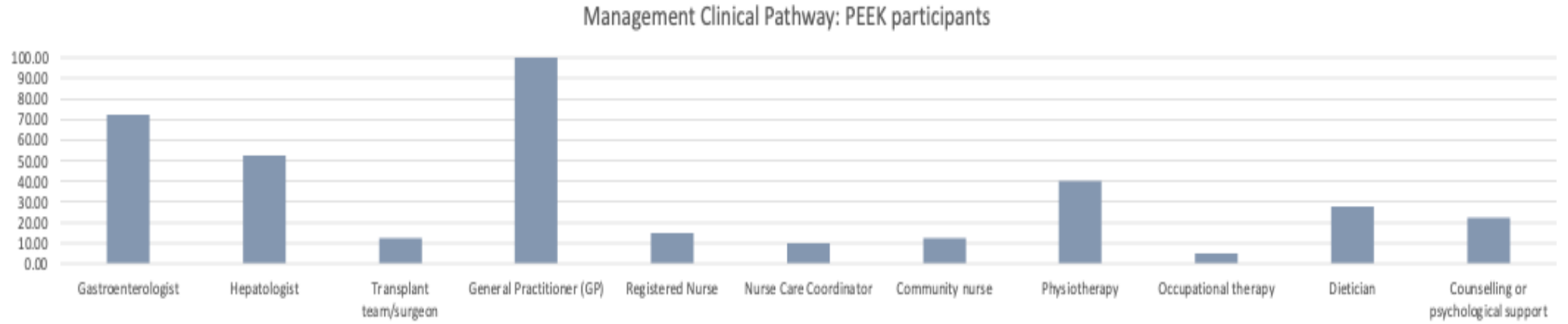
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Published clinical guideline recommendations

Category	Before 2020 *(EASL 2017; AASLD 2018)*	After 2020 *(AASLD 2022; APASL 2022; CSH 2021)*
Psychological Support	Recognize psychosocial impact of symptoms (fatigue, pruritus); encourage patient support groups; screen for depression/anxiety opportunistically; no structured mental health pathway specified.	Routine screening for anxiety, depression, sleep problems; structured referral to counselling, CBT-based approaches; multidisciplinary symptom management teams; patient education on symptom mechanisms; shared goal setting.
Lifestyle Modifications	Alcohol abstinence, smoking cessation, balanced diet with attention to fat-soluble vitamins in cholestasis; weight-bearing exercise for osteoporosis prevention; vaccination for hepatitis A/B if not immune.	Same core advice plus greater emphasis on personalized activity pacing, sleep hygiene, itch-relieving non-pharmacologic measures (e.g., emollients, avoiding heat), regular fracture risk assessment, periodic nutrition review, medication review for hepatotoxicity/pruritus worsening.

SUPPORTIVE CARE & EDUCATION

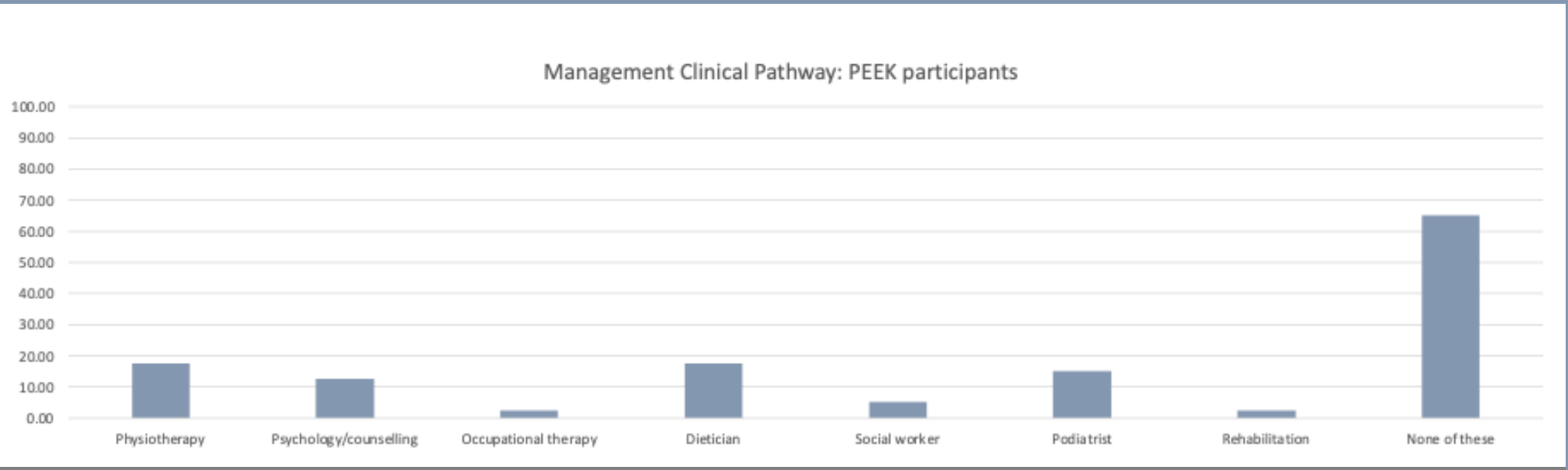
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Health professionals accessed

SUPPORTIVE CARE & EDUCATION

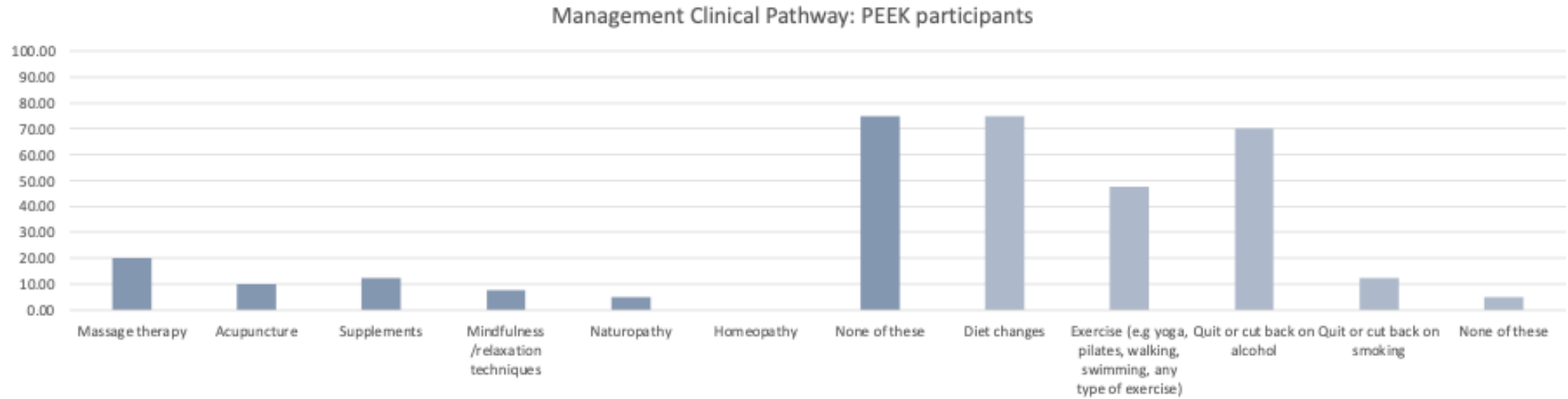
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Allied health accessed

SUPPORTIVE CARE & EDUCATION

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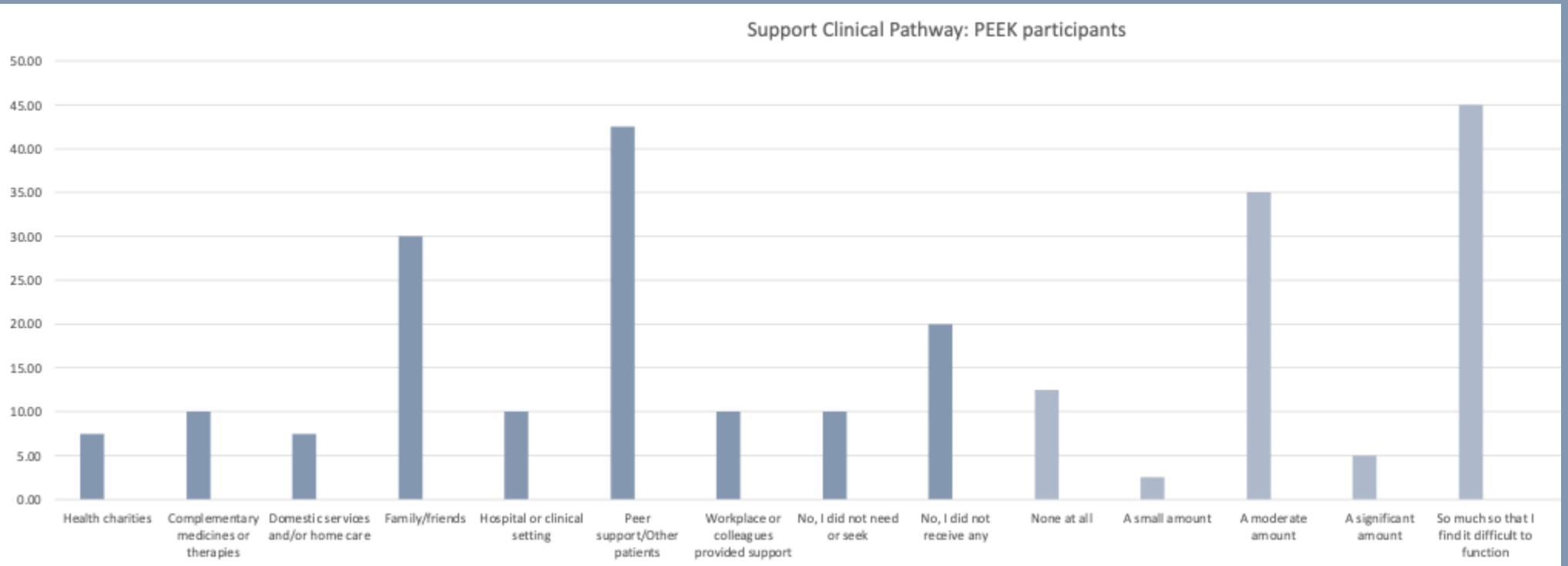


Complementary therapies accessed

Lifestyle changes

SUPPORTIVE CARE & EDUCATION

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Support accessed

Impact on emotional health

SUPPORTIVE CARE & EDUCATION

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Published clinical guideline recommendations before 2020

Long-term follow-up and monitoring

Before 2020, international guidelines from both the European Association for the Study of the Liver (EASL, 2017) and the American Association for the Study of Liver Diseases (AASLD, 2018) recommended lifelong follow-up for all patients with PBC, regardless of symptom status. The primary goals of monitoring were to assess biochemical response to ursodeoxycholic acid (UDCA), detect disease progression, and identify complications early. Biochemical markers—particularly alkaline phosphatase (ALP) and bilirubin—were advised to be checked every 3–6 months, as they are key prognostic indicators (EASL, 2017; Lindor et al., 2018). Additional monitoring included liver function tests, autoantibody status if initially negative, and thyroid function tests annually due to the association with autoimmune thyroid disease.

For advanced disease, guidelines recommended ultrasound surveillance for hepatocellular carcinoma (HCC) every 6 months in male patients and in women with established cirrhosis (EASL, 2017). Bone mineral density (BMD) assessment every 2–3 years was advised to monitor osteoporosis risk, alongside evaluation of fat-soluble vitamin levels in patients with advanced cholestasis. Patients with features of portal hypertension were to undergo endoscopic screening for varices at diagnosis of cirrhosis and at intervals based on baseline findings. The AASLD (2018) also emphasized the need for regular symptom assessment—including fatigue and pruritus—to guide supportive care, even when biochemical parameters are stable.

MONITORING & FOLLOW-UP

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Published clinical guideline recommendations after 2020

Long-term follow-up and monitoring

Recent guidance keeps lifelong follow-up centered on (1) monitoring biochemical response and prognosis, (2) detecting complications, and (3) symptom care. Panels recommend liver biochemistry every 3–6 months to gauge UDCA response/identify AIH features; use validated risk scores (GLOBE, UK-PBC) to refine long-term prognosis during follow-up; and consider non-invasive fibrosis tools as part of risk stratification (You et al., 2023; You et al., 2022). For cancer/portal-hypertension surveillance, perform ultrasound ± AFP every 6 months in cirrhosis and also in men with PBC; do upper endoscopy for varices in cirrhosis, repeating every 1–3 years based on findings and liver function (You et al., 2023). Extrahepatic monitoring includes annual thyroid testing, DEXA every 2–3 years (individualized to baseline BMD/cholestasis severity), and yearly fat-soluble vitamin levels when jaundiced (You et al., 2023). The AASLD 2021 update reinforces routine, structured symptom review (fatigue, pruritus, sleep/mood) and periodic reassessment with treatment escalation where needed (Lindor et al., 2022).

MONITORING & FOLLOW-UP

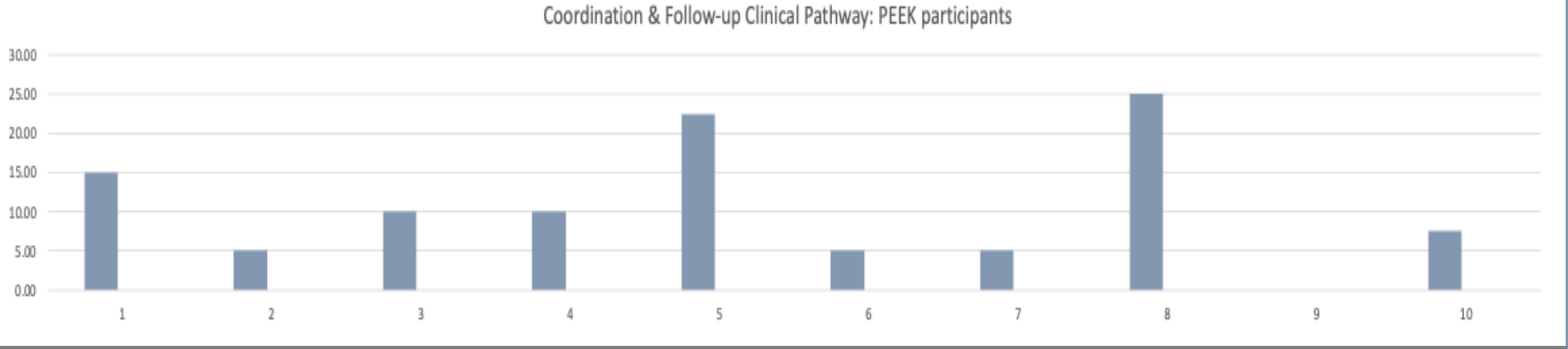
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Published clinical guideline recommendations

Monitoring Area	Before 2020 *(EASL 2017; AASLD 2018)*	After 2020 *(AASLD 2022; APASL 2022; CSH 2021)*
Biochemical monitoring	Liver function tests (esp. ALP, bilirubin) every 3–6 months to assess UDCA response; annual thyroid function tests; repeat AMA if initially negative.	Same 3–6 month liver biochemistry; annual thyroid tests; adds use of GLOBE and UK-PBC scores for prognosis; recommends non-invasive fibrosis tools for staging and follow-up.
HCC surveillance	Ultrasound ± AFP every 6 months in cirrhosis and in men with PBC.	Same groups; surveillance unchanged, but explicitly part of a structured follow-up algorithm.
Portal hypertension	Endoscopy at cirrhosis diagnosis, then every 1–3 years depending on variceal status.	Same timing; reaffirms intervals; includes elastography-based risk stratification in some regions.
Bone health	DEXA scan every 2–3 years; assess fracture risk; monitor fat-soluble vitamins in advanced cholestasis.	Same, but more emphasis on individualized DEXA interval based on baseline BMD; yearly vitamin level checks if jaundiced.
Symptom monitoring	Assess pruritus, fatigue at visits; no standardized tools required.	Structured symptom review at each visit; escalate therapy if symptoms worsen; may use validated PRO tools.

MONITORING & FOLLOW-UP

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Coordination received

Scale of 1 to 10, with 1 being poor and 10 being excellent

MONITORING & FOLLOW-UP

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PEEK
PRIMARY BILIARY CHOLANGITIS
AUSTRALIA 2025

Diagnosis

Timely access to diagnosis
- 50.00% Diagnostic test to result within 4 weeks

Diagnostic imaging and biopsy
- 36.00% Blood tests: Elevated ALP and GGT
- 32.00% Blood test: Positive AMA
- 25.00% Ultrasound
- 19.00% Liver Biopsy

Decision making

Health professionals are responsible for enabling patients to make informed choices according to their preferences, needs and values.
- 72.50% were offered only one treatment or approach

Treating specialists and practitioners should clearly explain the costs or how to find out the costs of services, tests and treatment options upfront
- 35.00%% were asked if they wanted to be treated as a public or private patient
- 57.50% were asked if they had private health insurance

Treatment

Treatment options
- 87.00%% had Ursodeoxycholic Acid (UDCA)

Clinical trials

Patients should be encouraged to participate in research or clinical trials where available and appropriate
- 10.00% discussed clinical trials with their doctor
- 82.50% were willing to take part in a clinical trial
- 5.00% participated in a clinical trial

Multidiscplinary teams

Multidisciplinary care involves all relevant health professionals discussing all relevant treatment options and making joint recommendations about treatment and supportive care plans, taking into account the personal preferences of patients. Main providers of care were:
- 40.00% Gastroenterologist
- 25.00% Hepatologist
- 25.00% General practitioner

Allied health

Supportive care is a vital part of any treatment program. It is made up of all the services, information and resources patients may need to meet their physical, psychological, social, information and spiritual needs from the time of diagnosis.
- 65.00% had no access to allied health professionals

Information

GPs and clinicians have responsibility for providing information to meet the physical, psychological, social, information and spiritual needs of patient
- 12.50% enough information at diagnosis
- 55.00% were given information about treatment options
- 52.50% were given information about managing their condition
- 32.50% were given information about diet
- 20.00% were given information about physical activity
- 17.50% were given information about psychological/social support

Communication

When anyone involved in treatment communicates with patients, they should be truthful and transparent but aware of cultural and psychological sensitivities.
- 80.00% had an overall positive experience of communication with healthcare professionals
- 70.00% described positive communication as two way, supportive and comprehensive conversations

45.00% understand their prognosis primarily meant it is a lifelong condition
- 20.00% had uncertainty around their prognosis

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Care coordination

Care coordination is the responsibility of every professional, both clinical and non-clinical, who works with patients, their families and carers.

5.21 was the average coordination of care score when rated on a scale of 1 to 10, with 1 being poor and 10 being excellent.

CCDR would like to thank each and every participant in this study