

A large, stylized orange grid graphic that resembles a globe or a sphere, composed of thick, curved lines that intersect to form a grid pattern. It is positioned in the background, partially obscured by a dark grey horizontal band at the bottom.

Navigating the management of cholestatic pruritus in patients with PBC: Insights from the multidisciplinary team

Practice aid for cholestatic pruritus in PBC

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Burden of cholestatic pruritus in patients with PBC



Occurs in up to **81% of patients** and may persist **chronically** in at least **35%**¹



Intensity of itch is **often worse at night**, causing **sleep deprivation, exhaustion** and **fatigue**^{1,2}



Pruritus shows inter-and intra-individual variation and is **not linked to PBC stage or severity**^{1,2}



Itch can have a **significantly detrimental effect on QoL** and can lead to suicidal ideation^{1,3}



No primary lesions or primary rash, though may see secondary lesions, e.g. excoriations, lichenification, prurigo nodules and scarring^{1,2}



Female patients report **more intense** pruritus **during hormonal changes**, e.g. the luteal phase of the menstrual cycle, during pregnancy or when taking HRT^{1,2}

Basic assessment of itch in PBC²

Patients may not associate itching with their PBC, so **may not proactively report their symptoms**

Cholestatic pruritus should, therefore, be assessed at the time of diagnosis and at all follow-up visits

Factors to assess include:



Intensity of itch



Time of start



Duration of itch



Localization of itch



Triggering factors



Relieving factors



Patient's opinion on the origin of itch



Burden of itch



Clinical examination of the entire skin to screen for primary and secondary skin lesions



Physical examination to rule out other pathologies that may be responsible for cholestatic pruritus

Patients may find it useful to **keep a record of pruritic activity in the form of a diary or in a digital format** that can be assessed together with their clinician at follow-up visits

Tools for assessing itch in PBC

NRS: intensity of itching is ranked from 0 (no itch) to 10 (worst itch imaginable)^{4,5}

VAS: intensity of itching marked on a 10 cm ruler (0=no itch; 10=worst itch imaginable)⁴

PGI-S: severity of itching at that time point is ranked from 1 (not present) to 7 (extremely severe)^{6,7}

PGI-C: change in severity of itching since baseline is ranked from 1 (very much improved) to 7 (very much worse)^{6,7}

5-D itch scale: five domains include **degree (severity)**, duration, direction, **disability** and distribution.⁷⁻⁹ First four domains measured on a 5-point Likert scale; 'distribution' includes 16 potential locations⁹

PBC-40: assesses **HRQoL** of patients with PBC with 40 questions over six domains (one of which is itch). Itch domain includes three questions to assess impact of itch over the last 4 weeks rated on a 5-point scale (never, rarely, sometimes, most of the time, always)¹⁰

Commonly used to measure pruritus at time of assessment or the worst pruritus in the previous 24 hours⁴

Treatment of cholestatic pruritus in PBC

EASL 2017 guideline recommendations¹¹

| Line of treatment | Agent | MoA | Approval |
|--|---|---|---------------------------|
| First-line | Cholestyramine | Bile acid sequestrant and anion exchange resin ^{2,11,12} | Yes ^{2,12} |
| Second-line | Rifampicin/rifampin¹³ | Antibiotic ^{12,14} | Off-label ^{2,12} |
| Third-line | Naltrexone or nalmefene | μ-opioid receptor antagonists ^{2,11,12} | Off-label ^{2,12} |
| Subsequent lines in unresponsive disease | Sertraline | SSRI | Off-label ^{2,12} |
| | Gabapentin | Anticonvulsant ¹⁵ | Off-label ² |



Liver transplantation when pruritus is 'persistent and intractable' after therapeutic trials¹¹

Japanese 2014 guideline recommendations¹⁴

| Line of treatment | Agent |
|-------------------|---|
| First-line | Cholestyramine |
| Subsequent lines | Rifampicin/rifampin¹³ |

Japanese approvals post-2014 guidelines^{2,12}

| Agent | MoA |
|--------------------|---------------------------|
| Nalfurafine | κ-opioid receptor agonist |

Practical tips for managing cholestatic pruritus

Patients should **receive education on pruritus as a symptom of PBC** as well as being advised on general **pruritus-relieving measures**²

Avoid skin dryness/irritation

- ✗ **Heat**, e.g. heavy or heat-retaining bedclothes^{2,17}
- ✗ Frequent (more than once a day) **washing with hot water**^{2,17}
- ✗ **Extensive rubbing** of the skin after showering²
- ✗ Contact with possible **irritants**, e.g. tea tree oil/chamomile²
- ✗ Overly **scented detergents**²
- ✗ **Tight clothing** or clothes made of animal wool^{2,16}
- ✗ Consumption of large amounts of **hot and/or spicy food, hot drinks or alcohol**²

Protect the skin and decrease pruritic activity

- ✓ Wash with **cold or lukewarm water**^{2,11,16,17}
- ✓ Use **mild/non-alkaline soaps and oils** that do not produce a lather^{2,17}
- ✓ Use **topical emollients** with cooling and/or anaesthetic effects (e.g. emollients containing **1–2% menthol or polidocanol**)^{2,11,16,17}
- ✓ Wear **soft, breathable clothing**^{2,17}
- ✓ Keep **nails short** to avoid skin damage^{2,17}
- ✓ Try **patting** rather than scratching/rubbing¹⁷
- ✓ **Manage stress**¹⁷

Some patients may benefit from relaxation techniques or psychological interventions for coping with the itch-scratch cycle²

Collaborating to support patients with PBC and cholestatic pruritus

- To improve disease outcomes, facilitate treatment adherence and increase patient QoL, **proactive pruritus management strategies should be implemented, including patient education and counselling**¹⁸
- Pruritus in PBC is subjective and carries individual threshold variations, therefore, **a comprehensive approach** to care is required.¹⁸ Management of patients with cholestatic pruritus should go beyond symptom monitoring and assume a **patient-centric attitude to all symptom management**¹⁸
- Signposting patients to **patient support groups** such as the [PBC foundation](#), or other national organizations can help them find support materials to help them in their daily lives¹⁹

Insights from a patient advocate

*“The itch in particular is very difficult to treat for many people, so I think **having the facility to listen to other patients, as well as doctors is very helpful.**”²⁰*



“Your hepatologist is your partner, you make decisions together, you discuss things together.”²⁰

Abbreviations and references

Abbreviations

EASL, European Association for the Study of the Liver; HRQoL, health-related QoL; HRT, hormone replacement therapy; MoA, mode of action; NRS, numerical rating scale; PBC, primary biliary cholangitis; PGI, Patient Global Impression; PGI-C, PGI-change; PGI-S, PGI-severity; QoL, quality of life; SSRI, selective serotonin reuptake inhibitor; VAS, visual analogue scale.

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