

A Standardized Clinical Assessment for Cholestatic Pruritus in Alagille Syndrome

DEAR HEALTH CARE PROVIDER,

As part of our shared commitment to positively impacting patients' lives, the Mirum Team strives to provide all the tools you need to provide a standardized assessment of patients with Alagille syndrome. The accompanying questionnaire can be used at every visit to monitor patients' progress.

In addition to this worksheet, you may refer to the Itch Severity Tool to help you and your patients have more meaningful, data-driven conversations about the itch and its impact. Patients can also download the Itch✓ app to track their itch between appointments.



Thank you for all you do for patients with Alagille syndrome. Together, we can help them and their families experience a brighter future.

Sincerely,

Your Team at Mirum Pharmaceuticals, Inc.



Assessing Patients With Cholestatic Pruritus in Alagille Syndrome

Today's date: Health care provider name:
(MM/DD/YYYY) *(First, Last)*

Patient name: Date of birth:
(First, Last) *(MM/DD/YYYY)*

Who's taking this survey today?
(Patient, parent, guardian, or someone else)

Review the following questions at each visit and select all that apply. If your patient is old enough to respond, please ask the questions to both the patient and caregiver.

1 What current medicine(s) and/or vitamin(s) are being taken? Have they changed recently?

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.....

2 How would you describe the itch on the worst days?



Not itchy
at all



Mild
(a little bit itchy)



Moderate
(somewhat/
moderately itchy)



Severe
(very itchy)



Very severe
(extremely itchy)

VISUAL ASSESSMENT

Clinician Scratch Scale (CSS) score¹:

Remember, patients may cover up their scratch marks so they aren't visible and may not actively scratch during their appointments. This doesn't mean they aren't experiencing unbearable itch. Use the following scale to assess your patients' itch severity today.

Score

0

Description

None

1

Rubbing or mild scratching when undistracted

2

Active scratching without abrasions

3

Abrasions

4

Cutaneous mutilation, hemorrhage, and scarring

Assessing Patients With Cholestatic Pruritus in Alagille Syndrome

3 How often does itching impact daily activities at school, work, and/or home?

- Every day
- 3 to 4 times per week
- Once or twice per week
- Once or twice per month
- Never

4 How often does itching make it difficult to fall asleep and/or stay asleep?

- Every day
- 3 to 4 times per week
- Once or twice per week
- Once or twice per month
- Never

5 With respect to treatment, how well are the following managed on a scale of 0 to 5? (0=not at all managed and 5=adequately managed)

- Weight
- Height
- Nutrition

Reference: 1. Gonzales E, Hardikar W, Stormon M, et al. *Lancet*. 2021;398(10311):1581-1592. doi:10.1016/S0140-6736(21)01256-3

6 Is there anything else to make note of today for your doctor?

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.....

.....

7 FOR CAREGIVER(S): Are you having trouble with sleeping and/or daily activities? Are there any other relevant updates you would like to share?

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HELPFUL TIPS FOR YOUR PATIENTS



Keep Tracking Symptoms Between Appointments

The Itch✓ app can be used twice a day to create a detailed journal of symptoms and observations, which can then be discussed at the next appointment.

Scan the QR code to learn more.

Grow Your Support System

The Alagille Syndrome Alliance (ALGSA) is a non-profit organization that supports the community by mobilizing resources, facilitating connections, promoting unity, and advocating for a cure.

Visit Alagille.org to get involved.



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