Member Name: {{MEMFIRST}} {{MEMLAST}} **DOB:** {{MEMBERDOB}} **PA Number:** {{PANUMBER}}

CVS caremark™ {{PANUMCODE}}

Bylvay

Prior Authorization Request

CVS Caremark administers the prescription benefit plan for the patient identified. This patient's benefit plan requires prior authorization for certain medications in order for the drug to be covered. To make an appropriate determination, providing the most accurate diagnosis for the use of the prescribed medication is necessary. **Please respond below and fax this form to CVS Caremark toll-free at 1-866-249-6155**. If you have questions regarding the prior authorization, please contact CVS Caremark at **1-866-814-5506.** For inquiries or questions related to the patient's eligibility, drug copay or medication delivery; please contact the Specialty Customer Care Team: CaremarkConnect® 1-800-237-2767.

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Patient's Name: {{MEMFIRST}} {{MEMLAST}} **Date**: {{TODAY}}
Patient's ID {{MEMBERID}} **Patient's Date of Birth:** {{MEMBERDOB}}
Physician's Name: {{PHYFIRST}} {{PHYLAST}}
Specialty: _____, NPI#: _____
Physician Office Telephone: {{PHYSICIANPHONE}} **Physician Office Fax:** {{PHYSICIANFAX}}
Request Initiated For: {{DRUGNAME}}

1. What is the diagnosis?
   - ❑ Pruritis in progressive familial intrahepatic cholestasis (PFIC)
   - ❑ Cholestatic pruritis in Alagille syndrome (ALGS)
   - ❑ Other _____
2. What is the ICD-10 code? _____
3. Has the patient received a liver transplant? ❑ Yes ❑ No
4. Is the requested drug being prescribed by or in consultation with a hepatologist or gastroenterologist?
   ❑ Yes ❑ No
5. Is the patient currently receiving treatment with the requested medication?
   ❑ Yes ❑ No *If No, skip to diagnosis section.*
6. Is the patient experiencing benefit from therapy (e.g., improvement in pruritis)? ***ACTION REQUIRED: If Yes, attach chart notes or medical records documenting a benefit from therapy (e.g., improvement in pruritis).***
   ❑ Yes ❑ No *No further questions.*

Complete the following section based on the patient's diagnosis, if applicable.

Section A: Pruritis in Progressive Familial Intrahepatic Cholestasis (PFIC)

7. Does the patient have progressive familial intrahepatic cholestasis (PFIC) type 2?
   ❑ Yes ❑ No *If No, skip to #9*
8. Does the patient have progressive familial intrahepatic cholestasis (PFIC) type 2 with variants in the ABCB11 gene resulting in non-functional or complete absence of the bile salt export pump protein?
   ❑ Yes ❑ No ***By answering No, you are attesting that the patient's genetic testing report does not indicate the presence of a variant that may be suggestive of nonfunctional BSEP-3 protein.***
9. What progressive familial intrahepatic cholestasis (PFIC) type does the patient have?
   ❑ PFIC type 1 ❑ PFIC type 2 ❑ PFIC type 3 ❑ Other _____

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10. Does the patient have a confirmed molecular diagnosis of progressive familial intrahepatic cholestasis (PFIC) type 1, 2, or 3? ***ACTION REQUIRED: If Yes, attach genetic testing results confirming a diagnosis of PFIC type 1, 2, or 3.*** ❑ Yes ❑ No

11. Does the patient have any other concomitant liver disease (e.g., biliary atresia, liver cancer, alternate non-PFIC related etiology of cholestasis)? ❑ Yes ❑ No

Section B: Cholestatic Pruritis in Alagille Syndrome (ALGS)

12. Does the patient have a diagnosis of Alagille syndrome (ALGS) established by genetic testing (e.g., mutations in JAG1 or NOTCH2 genes)? ***ACTION REQUIRED: If Yes, attach genetic testing results confirming a diagnosis of ALGS (e.g., mutations in JAG1 or NOTCH2 genes) and skip to #17.*** ❑ Yes ❑ No

13. Does the patient have a diagnosis of Alagille syndrome (ALGS) established by all of the following: A) family history of ALGS in a first degree relative, B) bile duct paucity, and C) one or more major clinical features of ALGS? *NOTE: Major clinical features of ALGS are: a) hepatic abnormality (e.g., cholestasis), b) cardiac abnormality (e.g., stenosis of the peripheral pulmonary artery and its branches), c) skeletal abnormality (e.g., butterfly vertebrae), d) ophthalmologic abnormality (e.g., posterior embryotoxon), e) characteristic facial features (e.g., triangular-shaped face with a broad forehead and a pointed chin, bulbous tip of the nose, deeply set eyes, and hypertelorism), f) central nervous system abnormality (e.g., stroke, intracranial bleeding), and g) renal structural or functional abnormality (e.g., abnormally small size, cysts). If Yes, skip to #17* ❑ Yes ❑ No

14. Does the patient have a diagnosis of Alagille syndrome (ALGS) established by both of the following: A) family history of ALGS in a first degree relative, and B) two or more major clinical features of ALGS? *NOTE: Major clinical features of ALGS are: a) hepatic abnormality (e.g., cholestasis), b) cardiac abnormality (e.g., stenosis of the peripheral pulmonary artery and its branches), c) skeletal abnormality (e.g., butterfly vertebrae), d) ophthalmologic abnormality (e.g., posterior embryotoxon), e) characteristic facial features (e.g., triangular-shaped face with a broad forehead and a pointed chin, bulbous tip of the nose, deeply set eyes, and hypertelorism), f) central nervous system abnormality (e.g., stroke, intracranial bleeding), and g) renal structural or functional abnormality (e.g., abnormally small size, cysts). If Yes, skip to #17* ❑ Yes ❑ No

15. Does the patient have a diagnosis of Alagille syndrome (ALGS) established by both of the following: A) bile duct paucity, and B) three or more major clinical features of ALGS? *NOTE: Major clinical features of ALGS are: a) hepatic abnormality (e.g., cholestasis), b) cardiac abnormality (e.g., stenosis of the peripheral pulmonary artery and its branches), c) skeletal abnormality (e.g., butterfly vertebrae), d) ophthalmologic abnormality (e.g., posterior embryotoxon), e) characteristic facial features (e.g., triangular-shaped face with a broad forehead and a pointed chin, bulbous tip of the nose, deeply set eyes, and hypertelorism), f) central nervous system abnormality (e.g., stroke, intracranial bleeding), and g) renal structural or functional abnormality (e.g., abnormally small size, cysts). If Yes, skip to #17* ❑ Yes ❑ No

16. Does the patient have a diagnosis of Alagille syndrome (ALGS) established by four or more major clinical features of ALGS? *NOTE: Major clinical features of ALGS are: a) hepatic abnormality (e.g., cholestasis), b) cardiac abnormality (e.g., stenosis of the peripheral pulmonary artery and its branches), c) skeletal abnormality (e.g., butterfly vertebrae), d) ophthalmologic abnormality (e.g., posterior embryotoxon), e) characteristic facial features (e.g., triangular-shaped face with a broad forehead and a pointed chin, bulbous tip of the nose, deeply set eyes, and hypertelorism), f) central nervous system abnormality (e.g., stroke, intracranial bleeding), and g) renal structural or functional abnormality (e.g., abnormally small size, cysts).* ❑ Yes ❑ No

17. Does the patient have evidence of cholestasis (e.g., elevated serum bile acid level)? ❑ Yes ❑ No

18. Does the patient have a history or presence of other concomitant liver disease (e.g., biliary atresia, progressive familial intrahepatic cholestasis [PFIC], liver cancer)? ❑ Yes ❑ No

I attest that this information is accurate and true, and that documentation supporting this information is available for review if requested by CVS Caremark or the benefit plan sponsor.

X_____

Prescriber or Authorized Signature **Date (mm/dd/yy)**

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