**Sample Format: Letter of Medical Necessity**

[Insert onto physician letterhead]

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| --- | --- |
| [Medical Director][Insurance Company][Address][City, State, ZIP] | **RE: Member Name** [Insert Member Name]**Member Number** [Insert Member Number]**Group Number** [Insert Group Number] |

**REQUEST:** Authorization for treatment with LIVMARLI® (maralixibat)

**DOSE AND FREQUENCY:** [Insert Dose & Frequency]

**REQUEST TYPE:** EXPEDITED/PRIORITY REVIEW

Dear [Insert Name of Medical Director]:

I am writing to support my request for an **expedited** **authorization** for my patient mentioned above to receive LIVMARLI® (maralixibat). LIVMARLI is an ileal bile acid transporter (IBAT) inhibitor indicated for the treatment of cholestatic pruritus in patients 12 months of age and older with progressive familial intrahepatic cholestasis (PFIC). Limitations of use: LIVMARLI is not recommended in a subgroup of PFIC type 2 patients with specific *ABCB11* variants resulting in non-functional or complete absence of bile salt export pump protein.

Progressive familial intrahepatic cholestasis is a rare, life-threatening disease, which can be diagnosed by clinical features or genetic test. There are many PFIC subtypes and additional ones are discovered as genetic testing of PFIC improves. The Online Mendelian Inheritance in Man (OMIM) compendium provides the most current understanding of PFIC subtypes. There are currently 13 known PFIC subtypes (list provided below). These subtypes may be referred to interchangeably by their PFIC subtype number, the affected gene, or by the protein deficiency if known.

PFIC presents in childhood with a range of clinical manifestations, including jaundice (yellowing of the skin), pruritus (itch), failure to thrive (impacted growth in height and weight), and progressive liver disease, which can lead to liver transplantation.

The cholestatic pruritus associated with PFIC is one of the most severe of any liver disease. The management of PFIC is challenging as there are limited therapeutic options to control pruritus. The only FDA approved treatments for cholestatic pruritus due to PFIC are IBAT inhibitors, which include LIVMARLI. Non-IBAT inhibitors are used off label and lack extensive clinical support for use.

LIVMARLI was studied in the broadest population of PFIC subtypes including some of those listed below. LIVMARLI showed meaningful improvements in cholestatic pruritis in just weeks and has a well established safety profile. It is also the only IBAT inhibitor with solid and liquid formulations.

This letter serves to document my patient’s diagnosis, medical history and to summarize my treatment rationale.

**Current Known PFIC Subtypes (OMIM)**

* **PFIC1**: OMIM #211600 – ATP8B1, AKA FIC1 deficiency or Byler disease
* **PFIC2**: OMIM #601847 – ABCB11, AKA BSEP deficiency
* **PFIC3**: OMIM #602347 – ABCB4, AKA MDR3 deficiency
* **PFIC4**: OMIM #615878 – TJP2
* **PFIC5**: OMIM #617049 – NR1H4
* **PFIC6**: OMIM #619484 – SLC51A
* **PFIC7**: OMIM #619658 – USP53
* **PFIC8**: OMIM #619662 – KIF12
* **PFIC9**: OMIM #619849 – ZFYVE19
* **PFIC10**: OMIM #619868 – MYO5B
* **PFIC11**: OMIM #619874 – SEMA7A
* **PFIC12**: OMIM #620010 – VPS33B – AKA Arthrogryposis, renal dysfunction, and cholestasis (ARC)
* **PFIC13**: OMIM #620962 – PSKH1

**Summary of Patient’s Diagnosis and History**

[Patient Name] is [Age] years old and was initially diagnosed with [Diagnosis] [ICD-10-CM] on [Date]. This diagnosis was confirmed by [insert details of patient’s genetic testing and PFIC subtype if known and/or clinical observations demonstrating PFIC]. [Patient Name] has been in my care since [Date].

[Insert a summary of the patient’s clinical history, current symptoms and condition, and relevant lab/test results (i.e. ALT, AST, TB, DB, INR, serum bile acid measurement, FSV). Highlight the factors leading you to recommend use of LIVMARLI and include any relevant previous treatments of pruritus with patient’s response to those interventions, such as Ursodiol, Rifampin, and antihistamines.

Note: Exercise your medical judgment and discretion when providing a diagnosis and characterization of the patient’s medical condition].

**Rationale for Treatment**

[Include your clinical rationale, patient’s likely prognosis without treatment with LIVMARLI and your credentials in treating PFIC].

Considering the patient’s history, condition, and the full Prescribing Information supporting uses of LIVMARLI, I believe treatment with LIVMARLI at this time is medically necessary and should be a covered treatment for my patient. [Include support for treatment rationale: You may consider including documents that provide additional clinical information to support the recommendation for LIVMARLI for this patient, such as the full Prescribing Information, peer-reviewed journal articles, or clinical guidelines].

Given the urgent nature of this request, please provide an expedited priority review and authorization. Contact my office at [insert phone number] if I can provide you with any additional information.

Sincerely,

[Insert Physician Name and Participating Provider Number]

Enclosures: [include full Prescribing Information and the additional support noted above].

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