

Continued Review of National Liver Review Board (NLRB) Guidance

OPTN Liver and Intestinal Organ Transplantation Committee

Purpose of Proposal

- The **National Liver Review Board (NLRB)** is responsible for reviewing model for end-stage liver disease (MELD) and pediatric end-stage liver disease (PELD) exception score requests
- The NLRB uses **OPTN guidance** to review exception cases that don't meet standardized criteria in OPTN policy
- Purpose:
 - Ensure **guidance remains updated, clear, and aligned** with current research so the appropriate candidates receive MELD or PELD exceptions

Proposal: Cystic Fibrosis Guidance

- CF is genetic disorder that can lead to chronic damage in the liver
- Current CF standardized exception policy aimed at lung-liver transplant candidates and is not pediatric specific
- Committee proposes new pediatric specific guidance for CFLD candidates meeting one of these criteria:
 - Portal hypertension with complications and patient has failed or is not a candidate for medical, endoscopic or surgical interventions to prevent or treat these complications
 - Growth failure as a result of liver disease, defined by age and sex-specific weight, length/height, weight-for-length, and/or body mass index (BMI) percentiles or moderate to severe malnutrition
 - Forced expiratory volume at 1 second (FEV_1) less than 70% or evidence of decline in FEV_1 of greater than or equal to 5% per year

Proposal: Hepatic Adenomas Guidance

- Hepatic adenomas (HA) are rare benign nodules occurring principally in women taking oral contraceptives
- Current NLRB guidance for multiple hepatic adenomas recommends candidates with HA and malignant transformation proven by biopsy or glycogen storage disease (GSD) be considered for a MELD exception
- Proposed changes:
 - Remove unnecessary intro paragraph to make guidance more succinct
 - Update criteria to better capture population of candidates needing MELD exception
 - Remove reference to “multiple” hepatic adenomas

Proposal: Budd-Chiari Syndrome

- Budd-Chiari syndrome is a medical condition characterized by hepatic vein thrombosis
- Proposed changes:
 - Remove unnecessary intro paragraph to make guidance more succinct
 - Add failed *surgical* management as qualifying criterion
 - Remove requirement for programs to provide etiology of hypercoagulable state
 - Remove the criterion related to decompensated hepatic hydrothorax requiring thoracentesis, which is already covered in guidance for hepatic hydrothorax

Rationale

- Cystic Fibrosis (CF) Guidance
 - Current policy does not consider pediatric-specific population
 - Adding pediatric specific guidance enables this population of candidates to access MELD or PELD exception scores
- Hepatic adenomas (HA) Guidance and Budd-Chiari Syndrome Guidance
 - Make guidance more succinct and clear
 - Update guidance to meet current clinical understanding

Member Actions

- Transplant programs and NLRB reviewers will need to be familiar with updated guidance

What do you think?

- The Committee is seeking public comment feedback on the proposed changes to NLRB guidance including:
 - The proposed creation of guidance for pediatric candidates with CF, specifically the proposed FEV1 thresholds set at less than 70% or greater than or equal to a 5% annual decline.
 - The proposed changes to guidance for candidates with hepatic adenomas.
 - The proposed changes to guidance for candidates with Budd Chiari.