Continued Review of National Liver Review Board (NLRB) Guidance

OPTN Liver and Intestinal Organ Transplantation Committee
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
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.