

**OPTN Liver and Intestinal Organ Transplantation Committee
National Liver Review Board (NLRB) Subcommittee
Meeting Summary
May 12, 2022
Conference Call**

James Trotter, MD, NLRB Chair

Introduction

The OPTN National Liver Review Board Subcommittee (the Subcommittee) met via Citrix GoToMeeting teleconference on 05/12/2022 to discuss the following agenda items:

1. Ongoing Review for NLRB Diagnoses

The following is a summary of the Committee's discussions.

1. Ongoing Review for NLRB Diagnoses: Guidance for Pediatric Cystic Fibrosis (CF) Candidates

The Subcommittee discussed updates to policy and guidance for Cystic Fibrosis (CF) candidates. The Subcommittee reviewed data for Hepatopulmonary Syndrome (HPS) and discussed updates to policy and guidance.

Summary of discussion:

CF:

The Vice Chair of the OPTN Pediatric Committee reviewed *OPTN Policy 9.5.B: Requirements for Cystic Fibrosis (CF) MELD or PELD Score Exceptions* and stated that the standard exceptions rarely apply to children on the liver transplant list. The OPTN Pediatric Committee Vice reviewed the drafted NLRB guidance for pediatric CF candidates for non-standard exceptions. To draft this section of the guidance, a group of pediatric subject matter experts met and reviewed available evidence to understand why the calculated MELD/PELD score does not accurately reflect waitlist mortality risk for pediatric transplant candidates who have CF-related liver disease (CFLD). The proposed guidance recommends that pediatric liver transplant candidates with CFLD should be considered for MELD/PELD exception points when any of the following criteria are met:

- Candidate has portal hypertension with complications, if the transplant program demonstrates that the patient has failed or is not a candidate for medical, endoscopic or surgical interventions to prevent or treat these complications.
- Candidate has growth failure, defined as age and sex-specific weight, length/height, weight-for-length, and/or BMI percentiles. Children and adolescents with CF and growth failure have a higher risk of waitlist mortality than children with non-CF related liver disease and therefore calculated MELD or PELD may not fully capture their risk of mortality.
- Candidate has an FEV1 < 70% or evidence of decline of > 5% per year, as these children may be expected to move toward advanced lung disease, reducing the opportunity for transplant.

A member asked if it was appropriate to include the criterion related to portal hypertension. The Pediatric Committee Vice Chair stated that there is already a section in the pediatric NLRB guidance which addresses portal hypertension complications, so this section was drafted as to not be redundant.

Another member suggested the portal hypertension section should remain because it addresses a specific population of patients. Staff stated that a reference could be included in this section.

Another member asked if there is guidance included for MELD score exceptions or if the proposed guidance is specific to PELD exceptions. The Pediatric Committee Vice Chair stated that the proposed guidance does not distinguish between MELD/PELD exceptions. The Pediatric Committee Vice Chair added that the guidance does not include a recommendation for a specific exception score because there was insufficient evidence to determine such a score.

A member asked whether guidance should include that transplant candidates should utilize CF transmembrane conductance regulator (CFTR) modulators first. The Pediatric Committee Vice Chair stated that the impact of CFTR modulators on liver disease is unclear in the long term. The Pediatric Committee Vice Chair stated that mentioning CFTR modulators in guidance may add confusion.

The NLRB Subcommittee supported the proposed guidance for pediatric candidates with CF.

HPS:

The Subcommittee reviewed *OPTN Policy 9.5.D: Requirements for Hepatopulmonary Syndrome (HPS) MELD or PELD Score Exceptions*. The Subcommittee reviewed data on candidates and recipients with an HPS diagnosis.

A member stated that no updates to policy or guidance are needed because the presented data did not show a significant difference in waitlist mortality for HPS transplant candidates. Other Committee members agreed.

The member added that it may be beneficial to review post-transplant outcomes data for transplant recipients with low PaO₂ at time of transplant. Members stated that their respective transplant programs use PaO₂ of 40 as a cutoff threshold. Another member suggested performing a literature review on this topic.

Next steps:

The Subcommittee will continue their review of this round of diagnoses during the next meeting.

Upcoming Meeting

- June 9, 2022 @ 2:30 PM ET (teleconference)

Attendance

- **Subcommittee Members**
 - Allison Kwong
 - James Trotter
 - John Lake
 - Kimberly Brown
 - Sophoclis Alexopoulos
- **HRSA Representatives**
 - Jim Bowman
 - Marilyn Levi
- **SRTR Staff**
 - Katie Audette
- **UNOS Staff**
 - Julia Foutz
 - Matt Cafarella
 - Susan Tlusty
- **Other Attendees**
 - Emily Perito