

**OPTN Liver and Intestinal Organ Transplantation Committee
National Liver Review Board (NLRB) Subcommittee
Meeting Summary
October 24, 2024
Conference Call**

James Pomposelli, MD, PhD, Chair

Introduction

The OPTN National Liver Review Board (the Subcommittee) met via WebEx teleconference on 10/24/2024 to discuss the following agenda item:

1. Project Development: Update NLRB Guidance + Further Alignment with Liver Imaging Reporting and Data Systems (LI-RADS)

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

1. Project Development: Update NLRB Guidance + Further Alignment with Liver Imaging Reporting and Data Systems (LI-RADS)

Presentation Summary

Hepatic Adenomas are benign tumors that can be singular or in clusters of multiple. Known risk factors include:

- Exposure to estrogens
- Anabolic steroids
- Genetic syndromes (Glycogen storage disease, Abernethy syndrome)
- Metabolic syndrome

Hepatic Adenomas come in three types:

- Hepatocyte nuclear factor (HNF)-1 alpha mutation
- Inflammatory
- Beta-catenin activation (malignant transformation)

Several factors determine treatment include patient symptoms, the size of the tumor, and the sex of the patient. Treatment options include observation, trans arterial embolization, radio frequency ablation, resection, and liver transplantation. The old guidance for Metabolic Disease did not have a priority score recommendation and tried to distinguish between metabolic disease with mild symptoms and with life threatening symptoms.

The suggested language changes to hepatic adenoma guidance include:

- an exception score of Median Model for End-Stage Liver Disease (MELD) at Transplant (MMaT)-3
- emphasis that the adenoma should be unresectable.
- include Abernethy malformation.
- change the language of the final three criteria to be unresectable, unresponsive, and has complications.

Summary of Discussion:

Decision #1: Added a line to Metabolic Disease guidance for the possibility of a higher exception score.

Decision #2: Reworded a line in Polycystic Liver Disease guidance.

Decision #3: Removed Portopulmonary Hypertension from guidance.

Decision #4: Removed a line from Early Allograft Dysfunction guidance.

Decision #5: Hepatic Hydrothorax guidance should be reformatted and add an exception score.

Decision #6: Hepatic Epithelioid Hemangioendothelioma guidance should be reformatted.

The Subcommittee had some concerns that the change to hepatic adenomas guidance stating “adenoma in the presence of underlying liver disease” may lead to unintended consequences where patients with a small adenoma may get transplant with little to no wait time which may not be fair relative to patients with more severe conditions like hepatocellular carcinoma (HCC). The Subcommittee discussed making the size of an adenoma a criterion as larger adenomas are more medically concerning and they also discussed changing this guidance specifically to glycogen storage disease or Abernethy malformation instead of underlying liver disease. By leaving the line underlying liver disease the Subcommittee was concerned that any patient with a cirrhotic liver and an adenoma could get MMat-3 when their lower MELD score is better representative of their priority.

The Subcommittee reviewed possible changes to the guidance for metabolic disease. They discussed adding a recommendation for MMat-3 for metabolic disease patients with mild symptoms and a MELD 40 score for patients that have metabolic disease with life threatening complications. The Subcommittee felt MELD 40 might be a bit extreme. They acknowledge that some patients may warrant more than MMat and decided to change the MELD 40 suggestion to say patients with life threatening complications may be considered for an increased priority score so that the guidance would preserve the possibility for higher exception scores when warranted.

The Subcommittee discussed the guidelines for Diffuse Ischemic Cholangiopathy. They debated on if the exception score should be set at MMat-3 or MMat. The suggested current change to guidance lists the exception score as MMat-3 but the current exception score is MMat. The Subcommittee felt they should continue to review that part of the suggestion later.

The Subcommittee discussed Primary/Secondary Sclerosing Cholangitis. The proposed change was to separate this guidance into two sections based on extremity of a patient’s condition. Less extreme cases were recommended for MMat-3, and more extreme cases were recommended for MMat. The Subcommittee felt it was best to try to match the language of this guidance to other guidance for consistency.

The Subcommittee noticed when reviewing Polycystic Liver Disease guidance that the first bullet point was already included in the introductory statement of the guidance and decided to reword the first line of the guidance for clarity.

The Subcommittee discussed Portopulmonary Hypertension being listed in guidance. This condition is already listed in policy so the Subcommittee debated if it should be removed or not. They considered leaving it as a reference point but felt it made more sense to remove it as there are other conditions that are in policy that don’t have references in guidance.

The Subcommittee discussed Early Allograft Dysfunction, formerly referred to as Reduced Size Syndrome and removed a line that said, “reduced sized livers” because they felt it was redundant.

The Subcommittee felt the guidance for Hepatic Hydrothorax suggestion was very detailed in part because it includes mortality data. They suggested taking it out for consistency as it would be easier to take the data out of this guidance than to add it into all the other guidance. They decided to put the data on risk and mortality at the bottom of the guidance for reference and to improve clarity. They also noted that the suggestion did not contain an exception score and agreed that MMaT-3 was appropriate.

The Subcommittee liked the suggested changes for Hepatic Epithelioid Hemangioendothelioma (HEHE) and felt it was appropriate guidance for the condition. They decided to reformat the suggestion to match other guidance by using bullet points to maintain consistency throughout the guidance documents.

Next steps:

- Guidance for hepatic adenomas will be reworked.
- HEHE and hepatic hydrothorax guidance will be reformatted.
- Decided on exception score for guidance on Diffuse Ischemic Cholangiopathy
- Submit updated guidance to the Liver and Intestines Committee for review.

Upcoming Meeting

- TBD

Attendance

- **Subcommittee Members**
 - James Pomposelli
 - Neil Shah
 - Joel Newman
 - Aaron Ahearn
 - Joseph DiNorcia
 - Michael Kriss
 - Cal Matsumoto
 - Allison Kwong
 - Shimul Shah

- **HRSA Representatives**
 - Jim Bowman

- **UNOS Staff**
 - Emily Ward
 - Cole Fox
 - Ben Schumacher
 - Niyati Upadhyay
 - Alex Carmack
 - Ethan Studenic