

Box 2: Weighing the Relative Risks and Benefits of Available Treatment Options

[2a] Parent participant (10001): *"We wanted to be in the driver's seat...for his best interest and his life...it's the same thing as a risk/benefit. You're making a pro and con list, and it's an unknown number of hyperammonemic episodes versus unknown complications from liver transplant. I think it's a hard decision, especially in those that might not be extremely sick right now."*

[2b] Provider participant (20011): *"Most challenging for the families – I think...they're probably, in many cases, wrestling that risk/benefit ratio. They're saying, 'How much chance do I want to take with my baby?' When we tell them how sick their baby can get from hyperammonemia, and then they hear from the surgeon how tricky the surgery could be... I see that as being a tough balance to accomplish."*

[2c] Provider participant (20001): *"I think the risk-benefit is much easier for families to see in kids that get transplanted for other things...where the kid is literally getting sick before your eyes in the hospital...we can tell you rather definitively that the only way your child is going to survive is to get a liver transplant as soon as possible. I think those decisions are just easier to make. The risk-benefit is much easier to assess because you can see how sick the kid is and how much medical support they're requiring... it can be much harder [in UCD] because the child looks okay and you're going along life as you've been doing and they have limitations but you get used to it. It's a really different decision, I think."*

[2d] Parent participant (10023): *"It just seems like – because there's no standard practice in these cases, it's extremely difficult. It feels like we're on the frontier of this thing, and it's not clear what the right decision is...we feel like we're making a decision, and we don't really have all of the information...Maybe that's just because that information doesn't exist."*