

Box 6: Disease Stability

[6a] Parent participant (10007): *“He was in the hospital every week or so because his ammonia would shoot up, and we really had no control over it. No matter what diet, how we adjusted his diet or his medications, his ammonia would always just go up. It was necessary for him to get a transplant because we couldn’t really control his ammonia.”*

[6b] Parent participant (10002): *“In her case, we just couldn’t keep her stable. She was pretty stable for two months before she was hospitalized again, but she had a handful of decompensations within the first few months. We were already feeling pretty desperate in talking about transplant within the first three months of her life”.*

[6d] Parent participant (10005): *“Initially, we were not for transplant. I, of course, did all the Google stuff, which I know you probably shouldn’t do, but I just saw all the complications and the constant taking of medication. It just didn’t seem like something that we wanted to do. We thought, oh, we can keep him managed, but basically, it started getting to the point where [he] was beginning to have to be hospitalized every couple of months for illness.”*

[6e] Parent participant (30002): *“From everything that I’ve heard, it should be for us more of a last resort scenario. My daughter’s condition, for the most part, has been pretty well-controlled. She’s had some, a few, maybe three or four high ammonia episodes in the last ten years that required hospitalization, and those were mostly well-controlled. In my mind, if that is still feasible for us, why would we take on the risks that are associated with the liver transplant?”*

[6f] Parent participant (10008): *“Tons of reasons as to why we felt transplant was a better option, even though he was quite a stable kid so far, but everybody told me that might not be the case forever. That can change literally overnight, so that’s why we decided to go for the transplant.”*

[6g] Parent participant (10031): *“She is stable. She’s met all her developmental milestones. She is thriving. You look at her, and she is very typical...So when she went into the [transplant] doctor’s office, they’re looking at us like, okay, she’s fine right now...but what about when she has her next one and...what is that going to look like? That terrifies us.”*