

Dear Reviewers:

Thank you for your valuable comments on the article, which is very important to improve the quality of the article. We have responded to the 3 comments you put forward and written them into the article:

1、 OTCD is a metabolic disease, protein diet restriction and medications that stimulate the removal of nitrogen from the body may restrain the progression of the disease. Unfortunately, these measures cannot prevent the emergence of hyperammonemia and metabolic encephalopathy, so liver transplantation is considered(**Add in TREATMENT**).

2、 Antibiotics prophylaxis (Sulbactam and Cefopcraxone) was used 8 d after surgery(**Add in OUTCOME AND FOLLOW-UP**).

3、 We have put arrow head at the biliary tracts in MRCP and intraoperative finding(**Add in FIGURE**).

Thank you again for your comments!