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ABO incompatible pediatric living donor liver transplantation

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ABO incompatible living donor liver transplantation (ABOi LDLT) has the potential to expand the donor pool for the patients with the end-stage liver diseases. There is a profound impact of recipient age on the incidence and severity of antibody-mediated rejection (AMR). Children younger than 1 year have low risks of AMR due to their immaturity of immune reaction. On the other hand, children aged 8 years or older have risks of hepatic necrosis; therefore, pretransplant desensitization by using rituximab is crucial to prevent AMR. At our institute, pretransplant desensitization with rituximab is used for children aged 1.5 years or older and it is given one month prior to their scheduled LDLT. In addition, if anti-donor ABO titer is more than x64, plasma exchange is introduced until it is lower than x8 before LDLT. Mycophenolate mofetil is also used as an additional immunosuppressant. One hundred twenty-one ABOi LT have been performed since 2010, and among them 27 patients (22.3%) received pretransplant desensitization. The median age of this group was 6 years. There were 4 patients with acute liver failure and 4 patients undergoing deceased donor LT, who did not receive rituximab at the appropriate timing before LT. Fifteen patients (12.4%) developed AMR in our ABOi series, including 6 patients without pretransplant desensitization and 9 patients with pretransplant desensitization. Cumulative incidence of AMR was 6.4% and 33.3% and the median onset of AMR was 11 days and 5 days after LT, respectively. All patients were successfully treated by steroid-pulse therapy, intravenous immunoglobulin (IVIG), and/or plasma exchange, although one patient at the age of 20 months without pretransplant desensitization suffered from intrahepatic cholangiopathy as a subsequent complication. In summary, ABOi pediatric LT shows excellent outcomes, although AMR related to ABOi LT cannot be completely eliminated. Treatment for AMR is not still standardized. Complete response is rarely achieved by conventional treatments, including plasma exchange, IVIG, steroids, or rituximab. New promising therapies for AMR are emerging.