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Sickle cell disease and liver transplantation: Review of 5 recent cases

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Position du problème et objectif(s) de l'étude:
Hepatic complications in sickle cell disease (SCD) are frequent. However, severe forms with terminal liver failure are rare. Liver transplantation (LT) then becomes a possible treatment. However, data regarding LT in patients with SCD are rather rare and limited to mostly case reports or small case series. We reviewed the last 5 patients who underwent a LT at Paul brousse in order to describe the characteristics of the patients at the time of their access to LT and their perioperative outcome

Matériel et méthodes:
Retrospective study of five patients. Results are expressed as number, percentage, mean ± standard deviation

Résultats & Discussion:
Patients had 37±16 years old (163±19 cm and 51±8 kg). The mean MELD score just before LT was 23±12. The main reason of their LT was acute liver failure on cirrhosis (N=3), fulminant hepatitis (N=1) and autoimmune hepatitis (N=1). All patients were hospitalized before their surgery (three on the ward and 2 in the intensive care unit). One patient with fulminant hepatitis was treated before surgery with MARS, and was under assisted mechanical ventilation and norepinehrine infusion. Surgical duration was 534 ± 35 minutes. Intraoperative transfusion was necessary in all patients with an average of 4±2 units of packed red blood cells. Two patients needed fresh frozen plasma and platelet transfusions. Intraoperative blood loss was 3180±2280 ml. Lactate concentration at the end of the surgery was 4.4±1.6 mmol/l.

Conclusion:
results: Only one patient had major hemodynamic instabilities intraoperatively. Norepinephrine at the end of surgery was on average at 0.7±0.2 mg/h. All patients (except one) were extubated on postoperative day#1. Intensive care unit and hospital length of stays were 7 ± 4 and 17 ± 2 days respectively. All patients presented at least one major postoperative complications (severe AKI requiring renal replacement therapy (RRT) (N=1), severe AKI not requiring RRT (N=2), pneumonia (N=1), biliary complication (N=1), portal thrombosis (N=1), retro-hepatic hemorrhage (N=1) , early allograft dysfunction (N=1), subarachnoid hemorrhage (N=1), multiple organ failure (N = 1). One patient died 16 days post-surgery from a subarachnoid hemorrhage.

Conclusion: LT for severe liver failure in sickle cell patients is feasible but is associated with an extremely high morbidity and mortality during the early postoperative period

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