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From Hepatic Failure to Recovery: A Child's Journey Through Liver Transplantation For Autoimmune Hepatitis - The Perioperative Pathway

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Abstract

Autoimmune hepatitis (AIH) is a chronic, progressive, immune-mediated hepatic disorder that can manifest as acute hepatic failure, particularly in paediatric and adolescent populations [1]. Despite significant advancements in diagnostic methodologies and immunosuppressive therapies, a subset of patients may experience rapid progression to sudden liver failure, necessitating urgent liver transplantation [2,3].

We present the case of an adolescent girl who exhibited acute hepatic failure attributable to autoimmune hepatitis. This condition was refractory to corticosteroid therapy and plasmapheresis, ultimately necessitating successful emergency liver transplantation. Her remarkable recovery underscores the critical significance of prompt recognition, timely referral, and a coordinated multidisciplinary approach to managing paediatric autoimmune hepatitis.

Key words: Autoimmune hepatitis, acute liver failure, paediatric liver transplantation, plasmapheresis.

Case Description

An adolescent previously healthy girl presented to our emergency department with a three-month history of jaundice, characterized by a distinctive yellowish discoloration of the sclera. She exhibited an abrupt onset of altered mental status and incoherent speech. Upon examination, she was profoundly icteric, lethargic, and exhibited mild hepatomegaly. Her vital signs were stable; however, she exhibited early manifestations of hepatic encephalopathy. Laboratory investigations revealed significantly elevated serum transaminases (AST/ALT), hyperbilirubinemia, a prolonged international normalized ratio (INR), and elevated serum immunoglobulin G (IgG) levels. The viral markers for hepatitis A, B, C, and E were negative. The autoimmune profile indicated positivity for antinuclear antibodies (ANA), smooth muscle antibodies (SMA), and a positive Coombs test, corroborating a diagnosis of Autoimmune Hepatitis Type I [4,5].

High-dose corticosteroid therapy, alongside three sessions of plasma exchange (PLEX) and one

session of continuous renal replacement therapy (CRRT), was commenced [6]; however, despite these intensive interventions, her clinical condition deteriorated, characterized by escalating bilirubin levels, exacerbating coagulopathy (INR >10), and progressive encephalopathy (stages II-III), ultimately satisfying the criteria for acute liver failure. Consequently, she was considered a candidate for orthotopic liver transplantation.

Since the patient presented with hepatic encephalopathy, she was electively intubated one day prior to the surgical intervention due to a diminished Glasgow Coma Scale (GCS) score. Antibiotic therapy, measures to mitigate coma, anticoagulants, and other supportive treatments were continued. All potential aspects of the procedure, including its advantages and disadvantages, as well as the necessity for liver transplantation as a definitive recovery option, were explained in detail to the family members. Following a comprehensive evaluation and Human Leukocyte Antigen (HLA) matching, her mother was designated as the donor.

On the day of the surgical procedure, a CT scan of the head and transcranial Doppler (TCD) ultrasound were performed on the morning of the procedure to exclude the possibility of irreversible cerebral injury. Acid aspiration prophylaxis was given in the preop holding area. Anesthesia was induced with a minimal dose of propofol, complemented by gradual administration of sevoflurane.

During the dissection phase (pre-anhepatic phase), the patient experienced significant haemorrhaging, accompanied by profound hypotension and tachycardia. Aggressive volume resuscitation using balanced crystalloids and blood product transfusions, meticulously guided by continuous hemodynamic monitoring and hourly arterial blood gas assessments, alongside bi-hourly laboratory evaluations, was administered. Numerous units of packed red blood cells (PRBCs) were transfused to maintain adequate oxygen-carrying capacity. Fresh frozen plasma (FFP) and prothrombin complex concentrate (PCC) were administered to replenish depleted coagulation factors and rectify the prolonged prothrombin time and elevated INR values, while platelet concentrates were administered to mitigate thrombocytopenia.

Calcium supplementation was administered to mitigate citrate-induced hypocalcemia resulting from massive transfusion, while concomitant infusions of antibiotics, albumin, antifungals, and N-acetylcysteine (NAC) were also administered. Normothermia was preserved through the use of fluid warmers and forced-air warming techniques to avert hypothermia-related coagulopathy. Metabolic acidosis was rectified through judicious ventilatory and fluid management complemented by bicarbonate supplementation. Invasive monitoring, including arterial pressure, FloTrac (for cardiac output assessment), and central venous pressure, facilitated precise volume replacement and vasoactive pharmacological support, including noradrenaline, adrenaline, and vasopressin infusions.

Inoperative point-of-care coagulation testing (TEG) was used to direct targeted blood component therapy. The International Normalized Ratio (INR) was elevated, and prothrombin complex concentrate (PCC) was initiated. Vasopressors, including norepinephrine, adrenaline, and vasopressin infusions, were administered to sustain the mean arterial pressure during the active haemorrhage episode. Adequate urine output was consistently preserved, indicating maintained renal perfusion. Following the effective implementation of transfusion therapy and hemodynamic optimization, hemostasis was achieved, and the patient was stabilized for progression to the anhepatic phase.

After complete mobilization, the hepatic inflow and outflow structures, including the hepatic artery, portal vein, and inferior vena cava, were occluded, leading to the explantation of the diseased liver. This signified the onset of the anhepatic phase, a pivotal interval during which the patient was temporarily devoid of hepatic metabolic and detoxifying functions. Clamping of the inferior vena cava precipitated a precipitous decline in venous return and cardiac output, resulting in hypotension, which was managed through volume resuscitation and administration of vasopressors (noradrenaline and vasopressin infusions). Metabolic acidosis and escalating lactate levels were addressed using

sodium bicarbonate and optimizing tissue perfusion. Hyperkalemia was rectified by infusing insulin and dextrose. Blood glucose levels were meticulously monitored every 60 min and maintained within a range of 120 to 180 mg/dL, utilizing dextrose infusion and insulin as necessary.

During this phase, meticulous attention was devoted to sustaining optimal preload and systemic vascular resistance, as hepatic venous clamping can precipitate hemodynamic instability. The ventilation was meticulously calibrated to ensure normocapnia and sufficient oxygenation. The absence of hepatic metabolism during this interval necessitates careful titration of anaesthetic agents to avert accumulation and protracted recovery. Coagulation parameters were diligently monitored and rectified with blood components guided by thromboelastography (TEG).

After completing the vascular anastomoses, the donor graft was reperfused through sequential unclamping of the portal vein and hepatic artery, signifying the commencement of the neohepatic or reperfusion phase. This phase was completed without any incidents. Prompt intervention involved the administration of intravenous calcium chloride and sodium bicarbonate and fluid resuscitation. The patient's hemodynamic status progressively improved as the graft commenced functioning. Continuous monitoring of electrocardiogram (ECG), invasive arterial pressure, entropy, temperature, cardiac output monitoring, arterial blood gases, pupillary reflex, urine output was done throughout.

After reperfusion, alprostadil infusion was initiated prior to hepatic artery anastomosis. Serial arterial blood gas analyses demonstrated a progressive amelioration of metabolic acidosis, which was indicative of graft functionality. The emergence of bile flow from the graft served as a reassuring indicator of viability. Coagulopathy, which arose from preexisting hepatic dysfunction and intraoperative hemorrhage, was adeptly managed through the judicious transfusion of fresh frozen plasma, cryoprecipitate, and platelets, guided by thromboelastography (TEG) and pertinent laboratory parameters. Calcium supplementation was administered as necessary. Blood glucose levels were meticulously monitored and regulated using insulin infusion. Normothermia was upheld, and urine output was ensured to exceed 0.5-1 ml/kg/hour, facilitated by meticulous fluid administration and intermittent furosemide as warranted.

At the conclusion of the surgical procedure, meticulous hemostasis was achieved, and all infusions were methodically tapered. The patient exhibited hemodynamic stability with minimal reliance on vasopressor support. Neuromuscular blockade was successfully reversed, and the patient was subsequently transferred to the intensive care unit for elective postoperative ventilation and rigorous monitoring of hemodynamic status, metabolic parameters, and graft function.

On the third postoperative day, a gradual weaning process commenced, during which the patient was successfully extubated following a satisfactory trial of continuous positive airway pressure (CPAP). Subsequently, the patient was maintained on high-flow nasal cannula (HFNC) oxygen therapy to ensure optimal oxygenation. The patient was gradually transitioned to oxygen via a nasal cannula and ultimately to ambient air. Early extubation in liver transplant recipients is correlated with enhanced pulmonary outcomes, reduced intensive care unit stay, and decreased postoperative complications.

Supportive management encompassed the administration of albumin to uphold oncotic pressure and intravascular volume, alongside blood transfusions to rectify anemia and enhance oxygen delivery. Meticulous regulation of electrolyte and fluid balance is imperative to avert graft congestion and renal impairment.

Immunosuppressive therapy was continued postoperatively, with tacrolimus dosages meticulously titrated according to serial serum concentrations and hepatic function parameters. Sustaining an optimal immunosuppressive milieu is paramount to thwart acute graft rejection while simultaneously

mitigating nephrotoxicity and other adverse effects.

The patient exhibited hemodynamic stability, satisfactory graft function, and progressive clinical improvement in the ensuing days. Early postoperative optimization of respiratory status, meticulous hemodynamic and laboratory monitoring, coupled with tailored immunosuppressive adjustments, were instrumental in realizing a favorable outcome.

The postoperative recovery was nothing short of extraordinary. Through the implementation of personalized immunosuppressive therapy and scrupulous multidisciplinary care, her hepatic function returned to normal within weeks. She regained full consciousness, appetite, and physical vitality, which was a remarkable transformation from critical hepatic failure to complete restoration.

Discussion

In a study of Rahim, et al.(2019) about the treatment of acute liver failure due to auto immune hepatitis particularly the decision between initiating corticosteroid therapy and proceeding to liver transplantation. They concluded that early corticosteroid therapy should be the initial treatment for most patients, as many will recover without requiring liver transplantation. Response to steroid must be assessed within 3-7 days. Patients who develop hepatic encephalopathy, coagulopathy or worsening of liver failure are unlikely to get benefit from steroid and should undergo early evaluation for transplantation. Delaying may worsen outcomes .

Another study of Hilberath J, et al (2024) about role of high volume plasmapheresis (HVP) in the management of pediatric acute liver failure concluded that high volume plasmapheresis can be a useful supportive therapy because it helps to remove inflammatory mediators and toxins while replacing deficient plasma components and thus leading to improvement in LFT, coagulopathy and hemodynamic instability. Hence, HVP should be considered as a bridge to either spontaneous liver recovery or liver transplantation.

In 2020 , Mittal R, et al .conducted a single center retrospective study and evaluated the outcomes of pediatric liver transplantation for auto immune liver diseases in 10 year period. Found that liver transplantation provides excellent patient and graft survival in children with ALF when medical therapy was unsuccessful, provided that patient is on good immunosuppressive therapy. Also careful long term follow up is essential to detect rejection, infection and recurrence of auto immune disease.

Maggiore G, et al. (2023) studied long term outcomes of patients with type 1 or type 2 auto immune hepatitis presenting in childhood [5]. The findings showed that with appropriate immunosuppressive therapy, long term overall survival is excellent, although a substantial proportion of patients requires liver transplantation. The 30 year overall survival was 81%, while native liver was 61%, indicating that many patients maintain their native liver for decades.

In our case the child was diagnosed with auto-immune hepatitis and was in acute liver failure with severe coagulopathy, which was refractory to corticosteroid and plasmapheresis, hence taken for emergency liver transplantation and successfully improved from the stormy pre operative and intraoperative period.

Conclusion

Autoimmune hepatitis can manifest dramatically in children and may progress to acute hepatic failure despite conventional treatment. Swift identification, early initiation of corticosteroid therapy, and timely liver transplantation are imperative for survival. We present this case report to underscore the critical importance of liver transplantation in reinstating hepatic function and enhancing the quality of life, particularly in the pediatric population. A multidisciplinary approach is essential to ensure optimal outcomes and foster long-term recovery, even in severe cases. Her convalescence

epitomizes the transition from turmoil to tranquility, a testament to both the resilience of the young patient and the advancements in modern hepatology, anesthesiology, and surgical techniques.

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