Case Report

Challenges in nursing care in achieving a successful outcome to a liver transplant on a 2-year old child with life threatening genetic condition

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Citation: Booma Ravi, Jaya Menon, Ruby Ravichandran, Farzana, Gayathri Devi. Challenges in nursing care in achieving a successful outcome to a liver transplant on a 2-year old child with life threatening genetic condition. Nightingale J., 2024;1(1):15-19.

Academic Editor: Dr. Venkita S. Suresh

1. Background

Liver Transplantation is successfully done for liver failure and liver cancer in selected patients. Liver transplantation for metabolic disorders due to genetic defect is rare and done in children mostly. When the disorder is severe and causes life threatening complications, they need to be done early, sometimes even less than a month after birth. The challenges in our case were that it was done for a metabolic disorder – Citrullinemia (urea cycle disorder), in a very small child (<10 kg). The urea cycle is a sequence of chemical reactions that takes place in the liver cells. These reactions process excess nitrogen that is generated when protein is used by the body. The excess nitrogen is used to make a compound called urea, which is excreted in urine.

Citrullinemia: Mutations in the ASS1 gene cause type I Citrullinemia. This gene provides instructions for making an enzyme, argininosuccinate synthase 1, that is responsible for one step of the urea cycle. Mutations in the ASS1 gene reduce the activity of the enzyme, which disrupts the urea cycle and prevents the body from processing nitrogen effectively. Excess nitrogen (in the form of ammonia) and other byproducts of the urea cycle accumulate in the bloodstream (hyperammonemia). Ammonia is particularly toxic to the nervous system, which helps explain the neurologic symptoms (such as lethargy, seizures, and ataxia) that are often seen in type I Citrullinemia.

This disease is inherited as an autosomal recessive genetic condition. Recessive genetic disorders occur when an individual inherits two copies of an abnormal gene for the same trait, one from each parent. If an individual receives one normal gene and one gene for the disease, the person will be a carrier for the disease and usually will not show symptoms. The risk for two carrier parents to both pass the defective gene and have an affected child is 25% with each pregnancy. The risk to have a child who is a carrier like the parents is 50% with each pregnancy. The chance for a child to receive normal genes from both parents and be genetically normal for that particular trait is 25%. The risk is the same for males and females.

2. Case presentation

A 1 year 8 months old female child, who had history of recurrent episodes of altered behavior (irritability) at 15 months of age, had presented to the casualty with altered sensorium. She was intubated and then started on intensive supportive measures. Her initial

workup showed features of hepatic failure. She had refractory seizures for which she was started on anti-epileptic. She underwent 2 sessions of plasma exchange. She showed clinical improvement, was extubated and started on oral feeds but then noticed to have waxing and waning of sensorium. Neuro imaging showed glutamine spikes. Metabolic work up showed elevated lactate, pyruvate, and citrulline. Provisional diagnosis of UCD (Urea Cycle Disorder) and Citrullinemia was considered. Genetic analysis confirmed the diagnosis of citrullinemia. She was started on low carbohydrate, low protein ketogenic diet and ammonia reducing measures, and continued on the anti-epileptic. With all above measures she remained stable and was on regular follow up. She was advised to undergo liver transplant to avoid further neurological complications.

She diagnosed to have urea cycle disorder with citrullinemia, and was evaluated for living donor liver transplant. Her mother volunteered and was accepted as the donor and completed her evaluation. The child was admitted for living donor liver transplant. She underwent a successful surgery with the left lateral lobe donated by her mother being transplanted to her. The graft sometimes needs to be reduced if there is any difficulty in the large graft placed in small babies, but no reduction was required for this baby.

The team was already prepared for the following challenges in the baby:

(1) Management of lines and tubes in a conscious baby

(2) Maintenance of double barrier nursing control to avoid infections as immunosuppression is started intra-operatively.

(3) Monitoring of vitals, intake -output closely every hour as the clinical situation can change in a matter of hours.

(4) Following up on all the investigations to collect the reports in a short turnaround time (TAT < 1 h) as the treatment is directed based on the reports.

(5) Nursing a small child for care and feeding in the absence of parents and relatives.

(6) Informing every issue of the child to doctors and following the orders promptly as it was very important.

Post-operative course

Living donor liver transplant - Left Lateral Segment done under GA

Post-transplant, child was shifted to intensive care unit. Child was hemodynamically stable. She was received with indwelling venous lines, arterial catheter and abdominal and urinary tubes. She was continued on mechanical ventilation in view of prolonged anesthesia.

POD 1: Baby extubated and was on nasal prong oxygen.

POD 2: Child developed breathing difficulty and tachycardia; she was found to have massive right sided pleural effusion. Re intubated and pigtail was inserted for pleural fluid drainage (265 ml, intermittently). Subsequently, oxygenation improved, tachycardia settled and procalcitonin showed a downward trend. Blood, pleural fluid and urine cultures, sent during this episode, were negative. Child gradually improved and was extubated on day 5. Doppler showed a normal post-transplant graft flow signal.

POD 7: Baby had one episode of melena and her Haemoglobin dropped. One unit of LDPRC started and PPI infusion also started. The anti-coagulation started to maintain vascular patency was also withheld.

POD 10: Child developed transaminitis that settled with appropriate change in immunosuppression and pulse dose of steroids (methylprednisolone 10 mg kg^{-1}).

POD 14: The abdominal drain output was feculent and child had tachycardia. Child underwent re-exploration, was found to have a fresh small bowel perforation and was repaired. Child was extubated the next day, after doppler showed a normal post-transplant study.

POD 18: Had fever, blood culture grew Klebsiella (pan sensitive) and antibiotic appropriated to cultures. Completed course of antibiotics. Follow up blood culture - sterile, procalcitonin normal.

The child gradually improved over the next few days with stable graft function and hemodynamics. Child was tolerating normal oral diet and was on 2 mg Tacrolimus at the time of discharge. Tac level 5.4 at the time of discharge. Other supportive medications and surgical prophylaxis were continued as per protocol. Steroid was tapered and tacrolimus dose was adjusted as per trough levels.

POD 24: Child was clinically well with good graft function and hence discharged on 03.01.2022

2.1. Nursing challenges in post-operative period

Respiratory concerns

Even though patient was extubated the next day, providing breathing exercise was the most challenging task. As the child was not blowing incentive spirometer, we provided sterile glove attached with small pipe to blow, which also was not accepted by the child. It was difficult to get the child to understand and co-operate. Hence, we encouraged the child to move left and right and allowed her to cry for sometimes to initiate breathing exercise.

When child developed respiratory distress, she was supported with HFNC.

Fluids and electrolytes imbalance

The baby had massive pleural effusion. Intake and output were carefully monitored and intake adjusted accordingly. TPN was adjusted and her glucose, potassium, proteins level monitored appropriately. The initiation of oral fluids was challenging as the baby developed paralytic ileus due to low potassium. Intravenous Inj. Potassium and Magnesium helped to reduce paralytic ileus, and gradually oral fluids were initiated to avoid intravascular fluid overload. Her blood sugar was elevated, so insulin infusion started. Because of insulin infusion further drop in potassium level was likely, hence adjusted carefully. While correcting potassium, it was diluted with 25% Dextrose to reduce the volume of total input and to maintain normal blood glucose.

Gastro-intestinal concerns

Pre-operatively baby was on low protein formula feed; baby refused to take high protein formula feed which was a challenging. It was gradually introduced, by 10 ml each time and increased to the maximum tolerated.

Medication management

Providing immune-suppressant medicine- Cap. Prograf- at 7 am – 7 pm was also challenging. The baby had to be kept nil per oral for an hour before and after the medication. Feeding schedules required co-operation from the baby which called for considerable rapport with the baby!

Ammonia level was maintained within normal limits

Administration of medication posed challenges: baby was on 12 regular medications (post-operative medicines not included) which included L-Arginine, Levocarnisure, Sodium Benzoate, Hepa-merz, antiepileptics, vitamins, calcium and Vitamin D3. On-time administration of medication orally was challenging and throughout the days in hospital stay. On-time administration of steroids, while maintaining normal glycemic level was required. On-time escalation and de-escalation of antibiotics according to cultures reports were called for. Appropriate usage of Albumin and TPN infusion required close monitoring to prevent fluid over load.

Hematology concerns

Blood and blood products were required to be administered to maintain Hb and coagulation profile.

Early identification of infection was done by proper monitoring of WBC and procalcitonin.

Acute rejection

Monitoring and on-time evaluation of acute rejection (temperature spike and elevation of SGOT and SGPT) and confirmation by doppler.

Proper administration of pulse steroids and adjustment of immunosuppressant was also challenging.

Care of tubes and drains

Conscious baby with more tubes and drains without sedation and without de-lining was also one of the great challenges.

Drained fluid-color and quantity were monitored and proper fluid replacement achieved, without overload.

Maintaining Central Venous Catheters (CVC)

Adherence to HICC protocol and proper handling of CVC were done. Proper administration of high alert medications and properly withdrawing blood for investigation.

Nephrotoxicity

Possible nephrotoxicity from the immunosuppressant (Tacrolimus) was managed by gradual adjustment of doses.

Vascular occlusion

Proper administration of anti-coagulant and anti-platelet aggregator was maintained to prevent vascular occlusion.

Early ambulation

Ambulated the baby, with tubes and drains.

Conclusion

Besides offering round the clock intensive nursing support, we were to also provide psychological and emotional support to a very young child who had a life-threatening disease and a life- defining surgery-liver transplantation.

Without the mother, managing a conscious baby by providing emotional support for 25 days was the greatest achievement, as we all were not nurses to the baby; we were mothers.