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Original Article

The impact of continuous renal replacement therapy for metabolic disorders in infants

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child

Background: While Continuous Renal Replacement Therapy (CRRT) is a well established treatment modality for patients with acute kidney insufficiency (AKI), it is now also being used for the management of various illnesses such as acute metabolic disorders presenting with hyperammonemia and elevated leucine levels. Herein, we aimed to describe our experience with CRRT in treatment of acute decompensation of 14 patients with a diagnosis of metabolic disorder who has been admitted to our pediatric intensive care unit (PICU) in the last year.
Methods: Patients who have had life threatening acute metabolic crisis due to various metabolic disorders and were treated with continuous renal replacement therapy (CRRT) were evaluated retrospectively.
Results: Between November 2014 and December 2015, 14 patients were found to have received CRRT for various metabolic disorders in the PICU. Ten patients had hyperammonemia and four patients had elevated leucine levels. Nine patients were male and five were female. The age interval was between 2 days and 18 months, with a mean of 5.5 ± 7.4 months. The weight distribution was between 2.5 and 18 kg, with a mean of 7.3 ± 5.6 kg. Eleven patients received continuous veno-venous hemodiafiltration (CVVHDF), and 3 patients with MSUD received continuous veno-venous hemodialysis (CVVHD). All patients have received high throughput hemodialysis and hemofiltration. The dialyzate rate was set to be minimum 4042 ml/h/1.73 m^2, and maximum 12,900 ml/h/1.73 m^2. Hemofiltration was performed with a replacement rate of 40—76 ml/kg/h. The average CRRT duration was 16.6 ± 15.6 h.

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Conclusions: We suggest that CRRT is an efficient method that can be used in hyperammonemia and elevated leucine levels which are metabolic emergencies.

1. Introduction

The recent renal replacement therapy (RRT) options consist of intermittent hemodialysis (IHD), peritoneal dialysis (PD) and continuous renal replacement therapy (CRRT). Although CRRT is accepted as a well-established treatment modality for patients with renal disorders such as acute kidney insufficiency (AKI), it is now also being used for the treatment of several non-renal indications. As a blood purification method, CRRT is indicated in hyperammonemia and elevated leucine levels which are considered as metabolic emergencies. Elevated ammonia levels are observed in metabolic disorders such as urea cycle disorders, organic acidemias and fatty acid oxidation defects. Ammonia is a well-known potent neurotoxin which may cause fulminant cerebral edema and death.[1] Urgent medical treatments should be applied to lower ammonia levels. However, medical treatment modalities do not act sufficiently quickly to lower very high ammonia levels. Thus, to prevent neurological interaction, RRT is recommended for ammonia levels over 400 μmol/L.[2]

Maple syrup urine disease (MSUD) is an inherited metabolic disorder characterized by organic acidemia due to the lack of branched chain ketoacid dehydrogenase complex. There are five sub-types and the most common type is the classical type. This type manifests as feeding difficulty, irritability, lethargy, rigidity, hypertonia, sometimes severe hypotonia, abnormal behavior and convulsions in the first two weeks of life.[3,4] In MSUD patients, the main neurotoxic agent is the amino acid leucine and the renal clearance of the circulating branched amino acids is low. If this condition is not treated as early as possible, it may cause cerebral edema and death. Thus, immediate clearance of excess leucine through RRT in severe metabolic crises is essential.[5]

In this study, we aimed to elucidate the role of CRRT as a safe and simple procedure in treatment of metabolic emergencies including MSUD.

2. Patients and methods

2.1. Study participants

Patients who received CRRT for metabolic coma in the PICU of Istanbul University Cerrahpasa Medical Faculty between November 2014 and December 2015 were examined retrospectively and patient information was gathered. CVVHD (continuous veno-venous hemodialysis) method was preferred for MSUD patients, while CVVHDF (continuous veno-venous hemodiafiltration) was the treatment of choice for other metabolic diseases presenting with hyperammonemia.

Age, sex, diagnosis, intensive care duration, catheter location, complications during and after CRRT, serum levels of toxic metabolites, infection rates and mortalities of the patients were examined.

2.2. Parenteral nutrition, supplementation and fluid infusion

A hypercaloric parenteral nutrition (110–130 kcal/kg/day) and intravenous thiamine supplementation were provided for patients with MSUD. To obtain an anabolic state, high dextrose containing fluid (16 mg/kg/sc) with slow insulin infusion 0.01–0.05 U/kg/min, lipid 3 g/kg/day and protein/BCAA-free formulas were provided.

For patients with hyperammonemia anabolic state was also provided. Arginine, citrulline, carboglu (carglumic acid) and sodium benzoate support were supplied. Also, after excluding the fatty acid oxidation disorders, intravenous lipid with 3 g/kg/day was started.

2.3. Catheterization and continuous renal replacement therapy

Femoral, internal jugular and subclavian veins were used for catheterization (SVC). Seven French (F) (Medcomp, Harleysville, Pennsylvania) hemodialysis catheters were used for two patients, while 8-Fr (Arrow; Arrow International, Reading, Pennsylvania, USA) double lumen hemodialysis catheters were used for the other patients. The catheter was dilatated by the dilatator of the 4-Fr catheter and then by dilatators of the 7-Fr and 8-Fr catheters. We used 4 Fr catheters as guide and used the smallest possible hemodialysis catheter.

Patients received sedation and analgesia prior to catheterization. Midazolam, ketamine, remifentanil were chosen for sedation and analgesia. Lidocaine was used as a local anesthetic. No neuromuscular blockers were used. No surgical catheters were attached. All catheters were attached by a single specialist.

Aseptic method was used during catheterization. The entry location was sterilized with 1% povidone-iodine and left to dry. Disposable sterile covers that covered the whole body were used. Sterile apron, mask and bonnet were used. Ultrasonography (USG) was not used. The catheters were attached with Seldinger method and fixed to the skin with 2.0 sharp-pointed silk sutures.

Prismaflex™ M60 (Gambro, USA) was used as a hemodialysis filter for 12 patients and Prismaflex™ HF20 (Gambro, USA) was used for two patients. All patients underwent