Therapeutic plasma exchange in pediatric patients of Guillain–Barre syndrome: Experience from a Tertiary Care Centre

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Abstract:

Background and Objective: Therapeutic Plasma Exchange (TPE) is performed effectively and safely in adult patients, but the use of TPE is limited in paediatric patients due to lack of universally accepted indications and technical challenges like establishment of adequate vascular access, low blood volume, increased incidence of adverse events during procedure and poor co-operation of patients during procedure. We present our experience of TPE in paediatric patients to assess the effectiveness and safety of TPE in paediatric patients. **Materials and Methods:** A total 122 TPE procedures were performed in 40 paediatric patients between 3 to 15 years of age group with Guillain Barre Syndrome (GBS). TPE procedures were performed on alternate days depending on the clinical condition of the patient. Patient's total blood volume was calculated as per Nadler's formula and processed through central double lumen catheter. 1-1.5 plasma volume was exchanged with normal saline and fresh frozen plasma. **Results:** A total of 122 TPE procedures were performed in 29 patients, of which 27 patients showed improvement from grade-0 and grade-1 to grade-III. One did not show any response and succumbed to the disease. Complication observed in 14 patients. **Conclusion:** TPE in paediatric patients has been increasing and has been shown to be effective as first line or adjunctive therapy in selected diseases. It is safe procedure when volume shifts, calcium supplementation and venous access are taken care.

Key words:

Guillain-Barre syndrome, pediatric patients, therapeutic plasma exchange

Introduction

Therapeutic plasma exchange (TPE) is a wellestablished modality of treatment in a variety of neurological, hematological, renal, and autoimmune diseases. It is performed effectively and safely in adult patients, but the use of TPE is limited in pediatric patients due to lack of universally accepted indications and technical challenges such as establishment of adequate vascular access, low blood volume, increased incidence of adverse events during the procedure, and poor co-operation of patients during the procedure.^[1-7] The decision to treat pediatric disease with apheresis is often based on the conclusions extrapolated from the adult patients; however, there has been reluctance to choose apheresis as a first line therapy in pediatric patients, even for conditions in which efficacy in adults has been conclusively proven because pathophysiology, clinical course, and therapeutic responses may differ in children.^[8] Nevertheless, the improvement in procedural techniques and the availability of various types of central venous catheters and ports have enhanced the safety and encouraged the use of apheresis in children. We present our experience of TPE in pediatric patients to assess the effectiveness and safety in pediatric patients.

Materials and Methods

In the present retrospective study, 40 patients with the age range of 3-15 years admitted to a tertiary care teaching hospital, from January 2013 to July 2014 who were referred to us for

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Correspondence to: Dr. Nidhi Bhatnagar, 2nd Floor, Kamdhenu Complex, Toran Dining Hall Lane, Opp. Sales India, Ashram Road, Ahmedabad - 380 009, Gujarat, India. E-mail: bhatnagarnidhi@ ymail.com performing TPE were evaluated. All patients had clinical findings of Guillain-Barre syndrome (GBS) and/or GBS variants. TPE procedures were performed on Spectra Optia apheresis machine (Manufacturer TERUMO BCT) on an alternate day using a double lumen femoral catheter depending on the clinical condition of the patient. A minimum of 1 and maximum of 5 cycles of plasma exchange were performed depending upon the clinical outcome in the patient. A total of 122 procedures were performed for 40 patients. Calcium gluconate infusion (10 ml of calcium gluconate in 500 ml normal saline [NS]) was given during the procedure to prevent citrate toxicity. Acid citrate dextrose: Whole blood ratio used was 1:10, blood flow rate was kept between 25 and 40 ml/min depending on the weight of the patient and blood volume of the patient was calculated. Depending on the amount of plasma exchange, the duration of procedure varied from 1 to 1.5 h. The TPE kit was primed first with NS and then with group specific, screened, and cross-matched packed red blood cells for patients who had weight <25 kg to avoid hypoxia and hypovolemia. Patient's total blood volume was calculated as per Nadler's formula and processed through central double lumen catheter. 1-1.5 times plasma volume was exchanged with NS and fresh frozen plasma (FFP) to prevent hypotension. The ratio of NS and FFP for replacement fluid was 1:1 [Table 1]. Continuous monitoring of vitals, e.g., pulse, blood pressure and respiratory rate was carried out during the procedure to prevent any adverse events related to the procedure. Details of the procedural complications, if any, were noted and analyzed. Pre- and post-procedure renal functions, coagulation parameter along with hematological parameters were done at every procedure. Clinical improvement was assessed by measuring the grading of muscle power as per Medical Research Council Scale.

Table 1: Average procedural data of TPE in pediatric patients

Age (in	Whole blood	Plasma	Replacement	ACD
years)	processed	volume	fluid transfused	used (ml)
	(ml)	removed (ml)	(ml)	
3-5	1137	515	571	93
6-8	1423	625	667	119
9-11	2009	961	927	170
2-15	2833	1398	1292	240

TPE: Therapeutic plasma exchange, ACD: Acid citrate dextrose

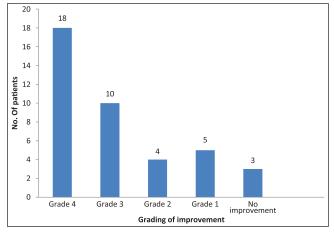


Figure 1: Clinical improvement after therapeutic plasma exchange in pediatric patients

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In our hospital, TPE is the first line of treatment for children of GBS weighing more than 15 kg. Intravenous immunoglobulin (IVIG) is preferable in children weighing <15 kg. Steroids are not used as a treatment protocol.

Inclusion criteria

Progressive weakness of variable degree from mild paresis to complete paralysis. Generalized hypo- or a-reflexia, demonstration of relative limb asymmetry regarding paresis. Mild to moderate sensory signs. Autonomic dysfunction include tachycardia, other arrhythmias, postural hypotension, hypertension, and other vasomotor symptoms. Gastrointestinal illness (e.g., diarrhea), cerebrospinal fluid (CSF) showing elevated CSF protein, CSF cell counts <10 mononuclear cell/mm³.

Results

A total of 122 TPE procedures (with an average of the three procedures per patient) were performed for 40 pediatric patients over a period of 1¹/₂ years from January 2013 to July 2014. In 29 patients, more than three TPE procedures were performed, out of which 27 patients showed improvement from grade-0 (complete paralysis) and grade-I (only a trace or flicker of movement in the muscle) to grade-III (movement possible against gravity but not against resistance) (Grading of muscle power is as per Medical Research Council Scale) [Figure 1]. One patient did not show any response and succumbed to the disease. Complications were observed in 14 patients who were well managed. Inadequate vascular access was most common complication observed in 11 patients [Figure 2]. It was due to inadequate flushing of the catheter by saline and heparin at regular intervals if unused. Other complications such as shivering, hypotension and machine error were also seen. The machine error was of centrifugation during the procedure, and it led to the abortion of procedure.

Discussion

The present study was done in the Department of Transfusion medicine of a tertiary care teaching hospital from January 2013 to July 2014. This study involved both male and female patients from 3 to 15 years of age groups. The main purpose of the study was to assess the efficacy and safety of TPE in pediatric patients. Most

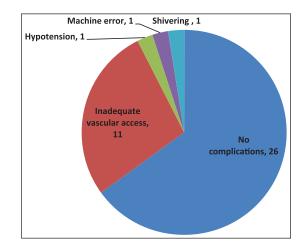


Figure 2: Complications of therapeutic plasma exchange in pediatric patients

of the patients underwent three cycles of exchange. The result of plasma exchange in terms of improvement in the clinical condition of the patient was excellent, that is, more than 85% in 18 patients. Of 40 patients, 18 patients' showed grade-IV improvement and 3 patients did not show any improvement. Totally, three patients were shifted to IVIG therapy when they failed to respond to TPE. No complication was seen in 26 patients, and minor complications were observed in 14 patients. The results of 29 pediatric patients in whom three or more TPE procedures were done, correlate well with previous studies. In our study, the most common complication of TPE was inadequate vascular access, but in a study conducted by Rekha Hans et al., the most common complication was allergic reactions to FFP. In a retrospective study by Michon et al. out of 186 children who had undergone a total of 1632 apheresis procedures, adverse reactions occurred in 55% of procedures, and 82% of patients.^[9] In this study, the most frequent complications were hypotension, symptomatic hypocalcemia, allergic reactions, and catheter-related adverse effects.

Conclusion

TPE can be performed safely in pediatric patients of GBS and has been shown to be effective as first line or adjunctive therapy in selected diseases. It shortens the course of hospitalization and reduces the mortality and incidence of permanent paralysis. It is a safe procedure when volume shifts, calcium supplementation, venous access; anticoagulation and psychosocial aspects are taken care of. Occasionally, minor complications such as hypotension and shivering occur.

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Conflicts of interest

There are no conflicts of interest.

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