New York Blood Center

Typical Uses for Apheresis in Emergent Situations

This document summarizes conditions for which apheresis, if indicated by the clinical situation, may be appropriate in emergent situations, to prevent or treat serious and life-threatening complications (1). This document does not purport to establish clinical standards, but is a summary of published literature on the subject (as indicated below), and is offered merely as an information source for NYBC Customers. Each treating physician should exercise his or her independent medical judgment as to the appropriate medical care for the presenting patient.

1. **Thrombotic thrombocytopenic purpura (TTP).** Emergent therapeutic plasma exchange (TPE) decreases the TTP mortality rate from >90% to <10% (2). Patients presenting with thrombocytopenia and microangiopathic hemolytic anemia alone may have TTP; the classic pentad (anemia, thrombocytopenia, fever, renal failure and neurologic changes) is uncommonly present. TTP may be associated with pregnancy, connective tissue disease, medications, and infection. Plasma infusion should be started while awaiting TPE(3). ADAMTS 13 activity should be drawn prior to the start of TPE. TPE should be performed on a daily basis until platelet and LDH normalization is present.

2. **Sickle cell disease (SCD).** Emergent red cell exchange (RCE) is used to treat life-threatening stroke, acute chest syndrome (ACS), or multi-organ failure. RCE, as compared to simple transfusion, is probably the optimal management for acute stroke and severe ACS (4). Although simple transfusion can be attempted in mild-moderate ACS, it risks increasing the hematocrit ≥ 30-33%, which increases blood viscosity and may exacerbate symptoms.

3. **Hyperviscosity due to Waldenstrom's macroglobulinemia (WM) or multiple myeloma (MM).** Emergent TPE is used to treat serious or life-threatening hyperviscosity symptoms such as bleeding, visual or hearing loss, seizures, and congestive heart failure. One procedure is typically needed. Treatment of underlying disease should also be initiated as soon as possible.

4. **Cytoreduction for leuko-, erythro-, or thrombo- stasis.** Emergent apheresis is used to treat cellular stasis due to leukemia and myeloproliferative diseases like polycythemia vera (PV) and essential thrombocythemia (ET). Symptoms and signs include neurologic symptoms, respiratory distress, gastrointestinal hemorrhage, and arterial/venous thromboembolic events. The target counts for cytoreduction are generally a WBC count < 50-100K/µl (AML) or < 400K/µl (ALL) with leukemias; a hematocrit < 45% with PV; and platelet count < 400-600K/µl in ET. These targets can usually be reached in 1-2 treatments.

5. **Rapidly progressive glomerulonephritis (RPGN).** Emergent therapeutic plasma exchange is used to treat ANCA-positive (Wegener's) RPGN who are dialysis-dependent at presentation; and anti-GBM (Goodpasture's) RPGN who are *not yet* dialysis-dependent, in order to decrease the risk of progression to end stage renal disease. Diffuse alveolar hemorrhage is also an indication

for urgent TPE in both variants. Treatment should continue until significant clinical improvement, which typically requires 6-10 procedures over 2-3 weeks.

6. **Antibody-mediated renal transplant rejection.** Emergent TPE is used when a kidney transplant recipient is experiencing acute humoral rejection, in order to increase the chance of graft survival. TPE should be instituted concomitant with immunosuppressive therapy, and typically involves daily to every other day treatments for 5-6 procedures.

7. **Myasthenia gravis crisis.** Emergent TPE is used to treat life-threatening respiratory compromise that could lead to intubation and aspiration. Importantly, TPE can benefit both antibody-positive and –negative patients, and may be more effective than IVIG in anti-MusK positive patients (5). A typical treatment course is daily to every other day procedures up to 2 weeks, with clinical improvement seen within 1-7 days.

8. **Wilson disease crisis.** Emergent TPE is used to treat fulminant hepatic and renal failure and hemolytic anemia. As a bridge to liver transplantation. TPE can rapidly decrease serum copper concentration to reduce progression of organ failure. After initial 1 - 2 procedures, further procedures are based on clinical/lab parameters.

1. Schwartz J*, et al.* (2013) Guidelines on the use of therapeutic apheresis in clinical practice-evidence-based approach from the writing committee of the american society for apheresis: the sixth special issue. *Journal of clinical apheresis* 28(3):145-284.
2. Bell WR, Braine HG, Ness PM, & Kickler TS (1991) Improved survival in thrombotic thrombocytopenic purpura-hemolytic uremic syndrome. Clinical experience in 108 patients. *The New England journal of medicine* 325(6):398-403.
3. Rock GA*, et al.* (1991) Comparison of plasma exchange with plasma infusion in the treatment of thrombotic thrombocytopenic purpura. Canadian Apheresis Study Group. *The New England journal of medicine* 325(6):393-397.
4. Swerdlow PS (2006) Red cell exchange in sickle cell disease. *Hematology / the Education Program of the American Society of Hematology. American Society of Hematology. Education Program*:48-53.
5. Pasnoor M*, et al.* (2010) Clinical findings in MuSK-antibody positive myasthenia gravis: a U.S. experience. *Muscle & nerve* 41(3):370-374.