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RMHP Apheresis, Therapeutic

AUTH: RMHP-A-017325 (AC)

MCG Health
Ambulatory Care
27th Edition

Link to Codes

Clinical Indications for Procedure

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Note: Prior authorization is not required for Photopheresis to treat Graft versus Host Disease (ICD D89.813). Because the CPT code is the same for the procedure, regardless of the diagnosis, the indication is included in the guideline. For claims processing purposes, do not void the case - no preauth. Rather, approve the request.

For members with **RMHP Medicare plans (CareAdvantage and Dual Special Needs Plan (DSNP)** plans, the case will be pended for the reviewer to apply the guidelines in the current National Coverage Determination (NCD) for Extracorporeal Photopheresis 110.4. See Reviewer Guidance.

Note: 0342T is not covered for any plan.

[**Expand All** / **Collapse All**]

- For **RMHP Individual and Family Plan (IFP) Commercial, PRIME (Medicaid) and CHP+** plan members, therapeutic apheresis may be indicated for **1 or more** of the following(1)(2)(3)(4)(5) :
  - Acute inflammatory demyelinating polyneuropathy (Guillain-Barre syndrome), as alternative to intravenous immunoglobulin(6)(7)
  - Antiphospholipid syndrome (catastrophic), as indicated by **ALL** of the following(8)(9):
    - Acute involvement of 3 or more organs, systems, or tissues
    - Antiphospholipid antibodies present

- Age-related macular degeneration (dry)
- Amanita mushroom poisoning

- Antiglomerular basement membrane disease, as indicated by **1 or more** of the following(10):
  - Diffuse alveolar hemorrhage
  - Patient not dialysis dependent, and creatinine less than 6.6 mg/dL (583 micromoles/L) [A]

- Antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis, as indicated by **ALL** of the following(12)(13):
  - Antineutrophil cytoplasmic antibody positive(12)
  - Appropriate clinical condition, as indicated by **1 or more** of the following:
    - Dialysis dependent
    - Dialysis is imminent.
    - Diffuse alveolar hemorrhage
- Autoimmune encephalitis(14)

- Babesiosis (severe), [B] as indicated by **1 or more** of the following(15):
  - Disseminated intravascular coagulation
  - Greater than 10% parasitemia
  - Pulmonary, renal, or hepatic dysfunction
  - Significant hemolysis (eg, blood hemoglobin level less than 10 g/dL (100 g/L), hemoglobinuria)

- Cardiac transplant, as indicated by **1 or more** of the following:
  - Cellular or recurrent rejection treatment needed
  - Desensitization prior to transplant
  - Rejection prophylaxis needed

- Chronic inflammatory demyelinating polyradiculoneuropathy, as indicated by **ALL** of the following(16)(17)(18):
  - Hyporeflexia or areflexia present in most limbs
  - Insufficient response to corticosteroids or intravenous immunoglobulin
  - Progressive or relapsing motor and sensory impairment of more than one limb

- Cryoglobulinemia, as indicated by **1 or more** of the following:
  - Membranoproliferative glomerulonephritis
  - Neuropathy (eg, mononeuritis multiplex)
  - Ulcerating purpura
  - Vasculitis(19)

- Focal segmental glomerulosclerosis, as indicated by **1 or more** of the following:
  - Post transplant: recurrent focal segmental glomerulosclerosis
  - Pretransplant: to prevent or delay recurrence
- Graft vs host disease, steroid-dependent or steroid-refractory
- Hemochromatosis (hereditary)

- Heterozygous familial hypercholesterolemia, as indicated by **1 or more** of the following(20)(21)(22)(23):
  - Patient with progressive coronary artery disease and **1 or more** of the following(24):
    - LDL cholesterol is greater than 200 mg/dL (5.18 mmol/L) or has decreased by less than 40% with medical therapy for 6 or more months.
    - Lipoprotein(a) is greater than 60 mg/dL (2.14 micromoles/L) and LDL cholesterol is greater than 125 mg/dL (3.24 mmol/L) despite medical therapy for 6 or more months.

- Patient without coronary artery disease and **ALL** of the following(24):
    - LDL cholesterol is greater than 300 mg/dL (7.77 mmol/L).
    - LDL cholesterol has decreased by less than 40% with medical therapy for 6 or more months.

- Homozygous familial hypercholesterolemia, as indicated by **ALL** of the following(20)(21)(25)(26):
    - Age is older than 2 years.
    - LDL cholesterol is greater than 500 mg/dL (12.95 mmol/L).(24)

- Hyperviscosity due to clonal thrombocytosis (eg, from essential thrombocythemia or other myeloproliferative disorder), as indicated by **1 or more** of the following:
    - Platelet count 1,500,000/mm^3 (1500 $x10^9$/L) or greater
    - Platelet count 450,000/mm^3 (450 $x10^9$/L) or greater and **1 or more** of the following:
        - History of thrombosis or bleeding
        - Vascular stasis signs or symptoms

- Hyperviscosity due to erythrocytosis, as indicated by **ALL** of the following:
    - Hematocrit greater than 55% (0.55)
    - Hyperviscosity symptoms
    - Simple phlebotomy has failed to reverse symptoms.

- Hyperviscosity due to leukocytosis, as indicated by **ALL** of the following(27):
    - Vascular stasis signs or symptoms
    - White blood cell count greater than 50,000/mm^3 (50 $x10^9$/L)

- Hyperviscosity [C] due to monoclonal gammopathy (eg, Waldenstrom macroglobulinemia, multiple myeloma with IgA, IgG, or kappa light chains), as indicated by **1 or more** of the following:
    - Neurologic signs or symptoms
    - Spontaneous bleeding from mucous membranes
    - Vascular stasis signs or symptoms
    - Visual disturbance due to retinopathy

- Lipoprotein(a) hyperlipoproteinemia, as indicated by **ALL** of the following:
    - LDL cholesterol is greater than 125 mg/dL (3.24 mmol/L) despite medical therapy for 6 or more months.
    - Lipoprotein(a) greater than 60 mg/dL (2.14 micromoles/L)
    - Progressive coronary artery disease
- Liver failure (acute)

- Liver transplant (ABO-incompatible), as indicated by **ALL** of the following(11):
    - Desensitization prior to transplant
    - Living related donor

- Lung allograft rejection, as indicated by **ALL** of the following:
    - Bronchiolitis obliterans syndrome
    - Failure of steroids or other immunosuppressive agents to halt syndrome progression
- Multiple sclerosis (acute, unresponsive to steroids)(29)(30)

- Myasthenia gravis, as indicated by **1 or more** of the following(31)(30)(32)(33):
    - During initiation of immunosuppressive therapy
    - During myasthenic crisis with ventilatory insufficiency or failure(34)(35)
    - During postoperative period after thymectomy
    - Prior to surgery (eg, thymectomy)(36)
    - Symptomatic patient resistant to or intolerant of immunosuppressive therapy(34)
- Mycosis fungoides (cutaneous T-cell lymphoma) for erythrodermic disease (stage III) [D]

- Neuromyelitis optica (acute), when high-dose intravenous steroids fail to resolve symptoms

- Pediatric autoimmune neuropsychiatric disorders associated with streptococcal infections (PANDAS), as indicated by **1 or more** of the following:
  - Refractory disease
  - Severe symptoms (eg, chorea, cognitive deficits, motor hyperactivity)

- Phytanic acid storage disease (Refsum disease), as indicated by **1 or more** of the following:
  - Acute neurologic or cardiac symptoms
  - Disease exacerbation
  - Maintenance therapy
- Polyarteritis nodosa associated with hepatitis B virus, in combination with glucocorticoids(38)(39)(40)
- Polyneuropathy due to monoclonal gammopathy (paraprotein neuropathy) with IgA, IgG, or IgM(41)

- Renal transplant (ABO-compatible), as indicated by **1 or more** of the following(42)(43):
  - Antibody-mediated rejection
  - Desensitization prior to transplant with crossmatch-positive living donor

- Renal transplant (ABO-incompatible), as indicated by **1 or more** of the following(42)(43)(44)(45)(46):
  - Antibody-mediated rejection
  - Desensitization prior to living donor transplant

- Sickle cell disease (acute) with complications, as indicated by **1 or more** of the following:
  - Acute stroke
  - Severe acute chest syndrome (ie, oxygen saturation less than 90% despite oxygen therapy)

- Sickle cell disease (nonacute) with complications, as indicated by **1 or more** of the following:
  - Cerebral infarct documented on brain MRI in absence of symptoms
  - High risk for stroke, as documented by transcranial Doppler study with mean blood flow velocity in the internal carotid artery or middle cerebral artery of 200 cm/second or higher
  - History of acute stroke or evidence of cerebral infarct on brain MRI
  - History of iron overload
- Thrombotic microangiopathy (drug-related)
- Thrombotic thrombocytopenic purpura(47)(48)(49)
- Vasculitis associated with HIV(39)
- Wilson disease

Alternatives to Procedure

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- Alternatives include conventional therapy for each condition.

Evidence Summary

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Background

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Therapeutic apheresis refers to extracorporeal technologies used to separate cellular (cytapheresis) and plasma (plasmapheresis) components of blood for a therapeutic purpose.(11)(50)(51)(52) **(EG 2)** Virtually any blood cell line can be effectively separated and removed from blood when its presence is pathogenic, as can dissolved plasma substances, such as immune complexes, cryoglobulins, toxins, myeloma light chains, and cholesterol-containing lipoproteins.(50)(51)(52)(53) **(EG 2)**

Criteria

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For acute inflammatory demyelinating polyneuropathy (Guillain-Barre syndrome), An expert consensus specialty society guideline states that there is high-quality evidence supporting the use of therapeutic plasma exchange as an alternative to intravenous immunoglobulin.(11)(30) **(EG 2)** As compared with supportive care alone, plasma exchange can accelerate motor recovery and reduce time on the ventilator; it is most effective when initiated within 7 days of the onset of symptoms.(11) **(EG 2)** A systematic review of randomized trials found moderate-quality evidence that plasma exchange significantly reduces time to recovery of motor activity. Although there was a significant increase in relapse in the first 6 to 12 months after treatment, the likelihood of full recovery increased and residual weakness decreased after 1 year.(7) **(EG 1)** A systematic review and meta-analysis found comparable efficacy and safety for plasma exchange and intravenous immunoglobulin for the management of Guillain-Barre syndrome.(32) **(EG 1)**

For catastrophic antiphospholipid syndrome, An expert consensus specialty society guideline supports the use of therapeutic plasma exchange as therapy for patients.(11) **(EG 2)** An international registry study evaluating outcomes following catastrophic antiphospholipid syndrome in 471 patients reported that mortality was 40.1% in patients treated with anticoagulation, cortico steroids, and either plasma exchange or intravenous immunoglobulin; mortality was 75% in patients who did not receive any of these treatments. There was no statistically significant difference in mortality between patients treated with plasma exchange and intravenous immunoglobulin, plasma exchange alone, or intravenous immunoglobulin alone.(54) **(EG 2)**

For age-related macular degeneration, An expert consensus specialty society guideline states that there is moderate-quality evidence supporting the use of rheopheresis (a form of plasmapheresis that separates out high molecular weight plasma components in order to reduce plasma viscosity) as therapy for this condition. Studies have demonstrated improvement in visual acuity lasting up to 4 years.(11) **(EG 2)** A randomized controlled study of 72 patients that evaluated the efficacy of rheohemapheresis found, at 12-month follow-up, that the treatment group had significant improvement in median best-corrected visual acuity, while the control group had a significant decrease.(55) **(EG 1)**

For Amanita mushroom poisoning, An expert consensus specialty society guideline supports the use of therapeutic plasma exchange for this condition. Patients who were treated with therapeutic plasma exchange were observed to have decreased mortality rates as compared with historical controls.(11) **(EG 2)**

For antiglomerular basement membrane disease, either with glomerulonephritis alone or with pulmonary hemorrhage (Goodpasture syndrome), An expert consensus specialty society guideline supports the use of therapeutic plasma exchange as therapy for patients who are dialysis independent or with diffuse alveolar hemorrhage.(11) **(EG 2)** Clinical trials and a single randomized controlled trial have demonstrated the effectiveness of apheresis, and total plasma exchange combined with prednisone and cyclophosphamide has become the standard of care.(53)(10)(56)(57) **(EG 2)**

For antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis (eg, granulomatosis with polyangiitis, microscopic polyarteritis), An expert consensus specialty society guideline supports the use of plasma exchange as therapy for patients who are dialysis dependent, who are in imminent need of dialysis, or who have diffuse alveolar hemorrhage.(11)(58) **(EG 2)** An open-label randomized controlled trial of 352 patients with severe, active, ANCA-associated vasculitis (treated with induction immunosuppressive therapy with either cyclophosphamide or rituximab) found that treatment with plasma exchange did not reduce the incidence of end-stage renal disease or death from any cause compared with placebo at 1-year follow-up. Study limitations included lack of renal biopsy documenting accurate kidney damage on entry to the study and wide confidence intervals for some

subgroups.(59) **(EG 1)** A systematic review and meta-analysis of interventions for renal vasculitis identified 8 studies investigating plasma exchange as adjunctive therapy in adults and found, at 3 and 12 months post treatment, that the intervention significantly reduced the need for dialysis.(60) **(EG 1)** An observational study of 137 patients with ANCA-associated vasculitis who were either on dialysis or had a serum creatinine greater than 5.6 mg/dL (495 micromoles/L) found, at median follow-up of 4 years, that there was no difference between treatment with plasma exchange or intravenous methylprednisolone with regard to the incidence of end-stage renal disease, relapse, and mortality. The authors noted that the study had limited statistical power to reliably conclude if there is a difference in clinical benefit for plasma exchange or intravenous methylprednisolone. Additional studies were recommended.(61) **(EG 2)** For children, a review article states that there have not been any trials of therapeutic plasma exchange for ANCA-mediated renal disease.(42) **(EG 2)**

For autoimmune encephalitis, Anti-N-methyl D-aspartate receptor (NMDAR) encephalitis is characterized by a prodrome of headache and fever, followed by sequential symptoms that may include behavioral changes, psychosis, catatonia, decreased level of consciousness, dyskinesias, and autonomic instability. Diagnosis is usually based on demonstration of NR1 IgG antibodies in serum or CSF, and initial therapy may include corticosteroids, intravenous immunoglobulin, or plasma exchange.(14) **(EG 2)** An expert consensus specialty society guideline supports the use of therapeutic plasma exchange for this condition, either as stand-alone therapy or in combination with other modalities (eg, intravenous immunoglobulin or high-dose methylprednisolone).(11) **(EG 2)** A systematic review of 71 studies (242 patients) evaluating plasma exchange in children (mean age 10.6 years) with anti-NMDAR encephalitis found a trend toward better outcomes when plasma exchange was administered with corticosteroids and started within a month of disease onset; however, the authors cautioned that these findings should be interpreted with caution given the limited number of patients and the retrospective nature of the studies.(62) **(EG 1)** Limbic encephalitis associated with voltage-gated potassium channel antibodies has been shown to respond within a few weeks to intense immunotherapy, which may include high-dose intravenous corticosteroids, intravenous immunoglobulin, or plasma exchange.(63) **(EG 2)**

For babesiosis, An expert consensus specialty society guideline states that red blood cell exchange is acceptable second-line therapy, either as stand-alone treatment or in combination with other modalities (eg, antibiotics) for patients with a severe form of this condition.(11) **(EG 2)**

For cardiac transplant, ABO-compatible, An expert consensus specialty society guideline states that there is high-quality evidence supporting the use of extracorporeal photopheresis for rejection prophylaxis and moderate-quality evidence for cellular or recurrent rejection. In combination with immunosuppressive agents, extracorporeal photopheresis has been shown to reduce both rejection episodes and allograft coronary artery vasculopathy. For pretransplant desensitization, therapeutic apheresis is an acceptable second-line therapy as a stand-alone treatment or in combination with other modalities (eg, intravenous immunoglobulin and rituximab).(11)(64) **(EG 2)**

For chronic inflammatory demyelinating polyradiculoneuropathy, An expert consensus specialty society guideline states that there is moderate-quality evidence supporting the use of therapeutic plasma exchange as therapy for this condition.(11)(30) **(EG 2)** A systematic review of randomized controlled trials found that there was moderate-quality to high-quality evidence from 2 small trials (44 patients) that plasma exchange provides significant short-term improvement in disability and clinical impairment scales, as well as nerve conduction velocity, but with the risk of rapid deterioration.(16) **(EG 1)** A combined analysis of a study of 30 childhood-onset chronic inflammatory demyelinating polyradiculoneuropathy patients and data from 11 previously published studies involving 113 patients reported a good response (improvement to the point of minimal to no functional impairment or limitation of activities, as per the treating physician) in only 14% of patients treated with plasma exchange, leading the authors to conclude that this therapy appears less useful for childhood chronic inflammatory demyelinating polyradiculoneuropathy.(65) **(EG 2)** Symptom relapse can occur after discontinuation of treatment.(11)(6) **(EG 2)** In randomized controlled trials, therapeutic plasma exchange, corticosteroids, and intravenous immunoglobulin achieve similar treatment outcomes.(11) **(EG 2)**

For cryoglobulinemia, An expert consensus specialty society guideline supports the use of therapeutic plasma exchange and immunoadsorption for this condition. Plasma exchange may be useful as adjunctive therapy for severe active disease characterized

by renal impairment, neuropathy, arthralgia, and/or ulcerating purpura.(11) **(EG 2)** Plasma exchange is also considered an effective therapy for vasculitis caused by cryoglobulinemia, including hepatitis C virus-related cryoglobulinemic vasculitis.(19)(66)(67) **(EG 2)**

For focal segmental glomerulosclerosis that is recurrent in a transplanted kidney, An expert consensus specialty society guideline states that there is moderate-quality evidence supporting the use of therapeutic plasma exchange as therapy for this condition. Patients with primary (etiology unknown) focal segmental glomerulosclerosis with proteinuria greater than 3 g/day do not benefit from therapeutic plasma exchange and should be treated with corticosteroids. For patients with secondary (eg, due to infection, toxins, drugs) focal segmental glomerulosclerosis, the underlying cause should be treated when possible.(11) **(EG 2)**

For graft vs host disease (acute or chronic), Evidence-based guidelines support the use of extracorporeal photopheresis for patients with steroid-dependent or steroid-refractory acute or chronic graft vs host disease. The most established role for extracorporeal photophoresis is in the treatment of steroid-refractory graft vs host disease, particularly with skin involvement. For nonskin manifestations, extracorporeal photophoresis has not been shown to be superior to alternative salvage therapies for acute graft vs host disease and may have a steroid-sparing effect in chronic graft vs host disease.(11)(68) **(EG 2)**

For hemochromatosis (hereditary), An expert consensus specialty society guideline states that there is moderate-quality evidence supporting the use of erythrocytapheresis as therapy for this condition.(11) **(EG 2)** A systematic review of 3 randomized trials (146 patients) found insufficient evidence to address how erythrocytapheresis compared with phlebotomy for the treatment of hereditary hemochromatosis; the authors noted that there is no evidence from randomized clinical trials that phlebotomy is of clinical benefit. The quality of evidence was low, and additional research was recommended.(69) **(EG 1)**

For heterozygous familial hypercholesterolemia, An expert consensus specialty society guideline states that there is high-quality evidence supporting the use of LDL apheresis as therapy for patients who are unresponsive to maximal medical therapy. A single treatment reduces LDL cholesterol by 65% to 70%, with demonstrated short-term improvements including improved myocardial and peripheral blood flow. Treatments are repeated indefinitely (usually every 1 to 2 weeks), with the frequency adjusted to reduce the time-averaged cholesterol by 60% or greater.(11) **(EG 2)** The Heart-UK LDL Apheresis Working Group recommends plasma exchange for patients with heterozygous familial hypercholesterolemia and progressive coronary artery disease when LDL cholesterol is greater than 200 mg/dL (5.18 mmol/L) or has decreased by less than 40% with medical therapy, or when lipoprotein(a) is greater than 60 mg/dL (2.14 micromoles/L) and LDL cholesterol is greater than 125 mg/dL (3.24 mmol/L) despite maximal medical therapy.(23) **(EG 2)** In the largest series published to date in a pediatric population with familial hypercholesterolemia followed for up to 21 years, LDL apheresis was found to be safe and effective in children as young as 3.5 years.(25) **(EG 2)**

For homozygous familial hypercholesterolemia, An expert consensus specialty society guideline states that there is high-quality evidence supporting the use of LDL apheresis as therapy for patients who are unresponsive to maximal medical therapy. A single treatment reduces LDL cholesterol by 65% to 70%, with demonstrated short-term improvements including improved myocardial and peripheral blood flow. Treatments are repeated indefinitely (usually every 1 to 2 weeks), with the frequency adjusted to reduce the time-averaged cholesterol by 60% or greater.(11) **(EG 2)** In the United States, it is generally accepted that patients with homozygous familial hypercholesterolemia who have LDL cholesterol levels greater than 500 mg/dL (12.95 mmol/L) are appropriate candidates for LDL apheresis.(70) **(EG 2)** A consensus practice guideline supports the use of LDL apheresis for patients with homozygous familial hypercholesterolemia, including children; treatment should be considered starting at 2 years of age.(71) **(EG 2)**

For hyperviscosity due to clonal thrombocytosis, An expert consensus specialty society guideline supports the use of thrombocytapheresis for symptomatic disease. The optimum role of thrombocytapheresis is not established for thromboprophylaxis or secondary thrombocytosis. Thrombocytapheresis has been utilized to prevent recurrent or treat acute thromboembolism or hemorrhage in patients with uncontrolled thrombocytosis and has been shown to be effective for microvascular ischemic complications that are not responsive to antiplatelet agents.(11) **(EG 2)**

For hyperviscosity due to erythrocytosis, An expert consensus specialty society guideline states that there is moderate-quality evidence supporting the use of erythrocytapheresis as therapy for this condition. For patients with polycythemia vera and acute thromboembolism, severe microvascular complications, or bleeding, erythropheresis can be used instead of large-volume phlebotomy, particularly if the patient is hemodynamically unstable.(11) **(EG 2)**

For hyperviscosity due to leukocytosis, An expert consensus specialty society guideline states that there is moderate-quality evidence supporting the use of leukocytapheresis as therapy for this condition. A single leukapheresis can reduce the circulating white blood cell count by 30% to 60%.(11) **(EG 2)** An observational study of 166 patients with acute myeloid leukemia and white blood cell count greater than 100,000/mm^3 (100 x10^9/L) with signs or symptoms of leukostasis compared no preinduction leukoreduction to leukoreduction modalities including leukapheresis, hydroxyurea, and leukapheresis and hydroxyurea and found , at median follow-up of 2.6 years, that there was no difference between groups with regard to 28-day mortality, complete remission, or overall survival.(72) **(EG 2)** A systematic review and meta-analysis of 21 studies (1354 patients) with acute myeloid leukemia and white blood cell count greater than 100,000/mm^3 (100 x10^9/L) comparing leukapheresis and hydroxyurea low-dose chemotherapy found that there was no significant difference between groups with regard to early mortality rate (deaths during first induction).(73) **(EG 1)**

For hyperviscosity due to monoclonal gammopathy, An expert consensus specialty society guideline states that there is moderate-quality evidence supporting the use of therapeutic plasmapheresis as therapy for this condition.(11) **(EG 2)** Another evidence-based guideline states that there is good evidence supporting the use of plasmapheresis for hyperviscosity syndrome due to Waldenstrom macroglobulinemia.(74) **(EG 1)** Plasmapheresis is effective in reducing the symptoms of hyperviscosity syndrome (eg, spontaneous bleeding from mucous membranes, visual disturbances due to retinopathy, neurologic impairment).(11)(28) **(EG 2)**

For lipoprotein(a) hyperlipoproteinemia, An expert consensus specialty society guideline states that there is moderate-quality evidence supporting the use of LDL apheresis for patients who are unresponsive to maximal medical therapy.(11) **(EG 2)** The Heart-UK LDL Apheresis Working Group recommends plasma exchange for patients with lipoprotein(a) greater than 60 mg/dL (2.14 micromoles/L) and progressive coronary artery disease when LDL cholesterol is greater than 125 mg/dL (3.24 mmol/L) despite maximal medical therapy.(23) **(EG 2)** A single-blind randomized sham-controlled trial of 20 patients with refractory angina and elevated lipoprotein(a) hyperlipoproteinemia (lipoprotein(a) level greater than 50 mg/dL (1.79 micromoles/L)) evaluating the efficacy of lipoprotein apheresis found, at 3-month follow-up, that apheresis was associated with improvement in quantitative myocardial perfusion reserve assessed by cardiovascular magnetic resonance. The authors recommended future larger studies to further evaluate this treatment, including patient selection and parameters.(75) **(EG 1)**

For liver failure (acute), An expert consensus specialty society guideline supports the use of high-volume therapeutic plasma exchange (defined as exchange of 15% of ideal body weight) for this condition; the role of therapeutic plasma exchange for this condition, however, is not established.(11) **(EG 2)** A multicenter randomized controlled trial of 182 adults with acute liver failure comparing standard medical therapy with or without high-volume plasma exchange found, after a mean of 2.4 treatments, that combined therapy was associated with higher survival to hospital discharge and transplant-free survival.(76) **(EG 1)**

For liver transplant (ABO-incompatible), An expert consensus specialty society guideline supports the use of therapeutic plasma exchange for desensitization prior to living donor transplant. The optimum role of therapeutic plasma exchange for desensitization prior to deceased donor transplant or humoral rejection is not established.(11) **(EG 2)**

For lung allograft rejection, An expert consensus specialty society guideline supports the use of extracorporeal photopheresis for this condition. The optimum role of therapeutic plasma exchange is not established for antibody-mediated rejection.(11) **(EG 2)**

For multiple sclerosis (unresponsive to steroids), An expert consensus specialty society guideline states that there is moderate-quality evidence supporting the use of therapeutic plasma exchange for multiple sclerosis patients with acute CNS inflammatory demyelinating disease that is unresponsive to steroids.(11)(77) **(EG 2)**

For myasthenia gravis, An expert consensus specialty society guideline supports the use of therapeutic plasma exchange as therapy for moderate to severe disease and prior to thymectomy.(11)(77) **(EG 2)** A systematic review and meta-analysis found comparable efficacy and safety for plasma exchange and intravenous immunoglobulin for the management of myasthenia gravis.(32) **(EG 1)** Prior to thymectomy, plasmapheresis has been shown to improve clinical outcomes, including less time on mechanical ventilation postoperatively and shorter ICU stay.(36) **(EG 2)** A cross-sectional analysis of 1053 myasthenia gravis patients showed that delaying plasma exchange for more than 2 days after admission was associated with higher mortality and higher complication rates than earlier treatment.(78) **(EG 2)** For myasthenia gravis resistant to or intolerant of immunosuppressive therapy, periodic therapeutic plasma exchange with 8-year follow-up in 11 myasthenia gravis patients who had frequent relapses on intravenous immunoglobulin showed that 9 have been in good control over the last 5 years, and the other 2 have lived normal lives without any treatment for the last 3 years.(34) **(EG 2)**

For mycosis fungoides (cutaneous T-cell lymphoma), An expert consensus specialty society guideline states that there is moderate-quality evidence supporting the use of extracorporeal photophoresis as therapy for erythrodermic disease. The role of extracorporeal photophoresis is not established for nonerythrodermic disease.(11) **(EG 2)** An expert consensus guideline recommends extracorporeal photophoresis for stage III disease with blood involvement.(79) **(EG 2)**

For neuromyelitis optica (acute), An expert consensus specialty society guideline states that there is moderate-quality evidence supporting the use of therapeutic plasma exchange for this condition. The role of therapeutic plasma exchange is not established for maintenance treatment.(11)(77) **(EG 2)**

For pediatric autoimmune neuropsychiatric disorders associated with streptococcal infections (PANDAS), An expert consensus specialty society guideline states that there is moderate-quality evidence supporting the use of therapeutic plasma exchange as therapy for this condition.(11)(77) **(EG 2)** Therapeutic plasma exchange has been shown to be effective in reducing symptom severity or shortening the duration of the disorder.(11)(30) **(EG 2)**

For phytanic acid storage disease, An expert consensus specialty society guideline supports the use of therapeutic plasma exchange and LDL apheresis for this condition.(11)(77) **(EG 2)**

For polyarteritis nodosa and other vasculitides, An expert consensus specialty society guideline supports the use of therapeutic plasma exchange as therapy for polyarteritis nodosa associated with hepatitis B virus in combination with glucocorticoids and antiviral agents; the role for therapeutic plasma exchange in idiopathic polyarteritis nodosa has not been established.(11) **(EG 2)** Systematic reviews of randomized controlled trials and expert opinion indicate that plasma exchange is effective in the treatment of hepatitis B-positive polyarteritis nodosa, HIV-associated vasculitis, and a variety of necrotizing systemic vasculitides.(39)(40) **(EG 2)**

For polyneuropathy due to monoclonal gammopathy, An expert consensus specialty society guideline supports the use of therapeutic plasma exchange as therapy for IgG, IgA, or IgM.(11)(77) **(EG 2)** Review articles conclude that randomized controlled trial data suggest effectiveness of plasmapheresis in improving neuropathy disability scores as well as summed compound muscle action potentials of motor nerves, especially in those with IgG or IgA gammopathy.(6)(41) **(EG 2)**

For renal transplant (ABO-compatible), An expert consensus specialty society guideline states that there is moderate-quality evidence supporting the use of therapeutic plasma exchange as therapy for antibody-mediated rejection or desensitization prior to transplant with a living donor. The optimum role of therapeutic plasma exchange for desensitization prior to transplant with a deceased donor is not established. For both antibody-mediated renal transplant rejection and desensitization, current regimens, which include plasma exchange, have a graft survival rate of up to 90%.(11) **(EG 2)**

For renal transplant (ABO-incompatible), An expert consensus specialty society guideline supports the use of therapeutic plasma exchange for desensitization prior to living donor transplant or antibody-mediated rejection.(11) **(EG 2)** Protocols that include

therapeutic apheresis have been associated with allograft and patient survival of greater than 95% at 18 months.(46) **(EG 2)** Five-year graft survival has been as high as 78%.(53)(31) **(EG 2)** A matched controlled study of renal transplant candidates with donor-specific human leukocyte antigen antibodies who underwent plasmapheresis and intravenous gammaglobulin desensitization before kidney transplant found that, as compared with waiting for a compatible organ, desensitization was associated with increased estimated patient survival.(45) **(EG 1)**

For sickle cell disease (acute), An expert consensus specialty society guideline and specialty society guidelines support the use of red blood cell exchange for the complications of acute stroke and severe acute chest syndrome. The role of red blood cell exchange is not established for complications of priapism, multiorgan failure, splenic/hepatic sequestration, and intrahepatic cholestasis. For complications of acute ischemic stroke and rapidly progressing acute chest syndrome, red blood cell exchange is preferred to transfusion, as the hemoglobin S (HbS) concentration is rapidly reduced by replacing red blood cells containing HbS with normal red blood cells, without causing volume overload.(11)(80)(81) **(EG 2)**

For sickle cell disease (nonacute), An expert consensus specialty society guideline supports the use of red blood cell exchange for stroke prophylaxis or prevention of iron overload in patients with sickle cell disease. Red blood cell exchange can remove or keep iron stores steady.(11) **(EG 2)** A randomized controlled study of 196 children (mean age 10 years) with sickle cell disease, no history of stroke, and one or more silent cerebral infarcts on MRI compared the efficacy of standard care vs regular red cell transfusions to maintain a target hemoglobin concentration. At a median follow-up of 3 years, transfusion was associated with a lower incidence of stroke or new or enlarged silent infarct.(82) **(EG 1)** A randomized controlled trial of 130 children with sickle cell disease who had no history of stroke but were considered to be at high risk due to abnormal results on transcranial Doppler ultrasonography compared treatment with transfusion (exchange or simple transfusion) vs standard care. At a median follow-up of 21 months, there was one stroke in the transfusion group compared with 10 strokes in the standard care group; the trial was stopped prematurely based on an interim analysis of the data.(83) **(EG 1)** A retrospective study evaluating the efficacy of long-term erythrocytapheresis (LTE) in 36 pediatric sickle cell patients found, after a mean of 5 years of therapy, improved growth and peak height velocity in the long-term erythrocytapheresis group compared with matched controls without evidence of complications related to iron overload.(84) **(EG 2)**

For thrombotic microangiopathy (drug-related), An expert consensus specialty society guideline supports the use of therapeutic plasma exchange as therapy for ticlopidine-associated disease. However, the role of therapeutic plasma exchange is not established for clopidogrel-associated, cyclosporine-associated, or tacrolimus-associated disease, and it is considered ineffective for treatment of disease related to gemcitabine and quinine.(11) **(EG 2)**

For thrombotic thrombocytopenic purpura, An expert consensus specialty society guideline states that there is high-quality evidence supporting the use of therapeutic plasma exchange as therapy for this condition.(11)(47)(48) **(EG 2)** An audit of 38 patients with this condition over 14 years showed that 82% achieved initial complete response with plasma exchange with fresh-frozen plasma and steroids; 50% of patients achieved sustained complete response, which was defined as no documented relapse for a minimum of 12 months.(49) **(EG 2)**

For vasculitis associated with HIV, Plasma exchange is able to rapidly clear the immune complexes responsible for the disease. Even though it may be possible to obtain good clinical results with antiviral drugs alone, the severity of the disease in most patients requires a regimen that can immediately control severe or life-threatening manifestations.(39) **(EG 2)**

For Wilson disease, An expert consensus specialty society guideline supports the use of therapeutic plasma exchange as therapy for this condition. The most common use of therapeutic plasma exchange is as a bridge to transplant for fulminant hepatic failure when a donor liver is not readily available.(11) **(EG 2)** An evidence-based pediatric practice guideline states that plasma exchange coupled with chelation therapy has demonstrated variable short-term improvement for children with Wilson disease who present with acute hemolytic crisis.(85) **(EG 2)**

Inconclusive or Non-Supportive Evidence

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For ABO-incompatible bone marrow or stem cell transplant, A study of 26 patients who underwent plasma exchange prior to ABO-incompatible bone marrow transplant found that, as compared with an ABO-identical donor group at 100 days, there was no significant difference in graft vs host disease, median time to engraftment, transfusion requirement, and treatment-related mortality. At median follow-up of 17 months, there was no significant difference in survival between the groups (38% for ABO-incompatible and 44% for ABO-identical).(86) **(EG 2)** A study evaluating the efficacy of antidonor isoagglutinin reduction prior to ABO-incompatible stem cell transplant using plasmapheresis (6 patients), transfusion of ABO-incompatible donor type red blood cells (70 patients), or combined methods (22 patients) found that prior isoagglutinin reduction significantly reduced incidence of post-transplant red cell aplasia as well as mean time to red cell engraftment.(87) **(EG 2)** An expert consensus specialty society guideline states that stem cell transplant studies demonstrate inconclusive results with regard to the use of pretransplant therapeutic plasma exchange on reduction of isoagglutinin titers, impact on time to red cell engraftment, and incidence of post-stem cell transplant pure red cell aplasia.(11) **(EG 2)**

For amyloidosis, An expert consensus specialty society guideline states that therapeutic plasma exchange provides no objective benefit for this condition.(11) **(EG 2)**

For amyotrophic lateral sclerosis, Several small studies of plasma exchange therapy failed to demonstrate substantial alteration in the course of disease.(88) **(EG 2)** An expert consensus specialty society guideline states that therapeutic plasma exchange provides no objective benefit for this condition.(11) **(EG 2)**

For aplastic anemia or acquired pure red cell aplasia, An expert consensus specialty society guideline states that the optimum role of therapeutic plasma exchange is not established for this condition.(11) **(EG 2)**

For atopic dermatitis, recalcitrant, An expert consensus specialty society guideline states that the roles of therapeutic plasma exchange, extracorporeal photopheresis, and immunoadsorption are not established for this condition.(11) **(EG 2)**

For autoimmune hemolytic anemia, An expert consensus specialty society guideline states that there is low-quality or very low-quality evidence supporting the use of therapeutic plasma exchange for cold agglutinin disease and warm autoimmune hemolytic anemia.(11) **(EG 2)**

For Behcet disease, An expert consensus specialty society guideline states that the role of therapeutic plasma exchange is not established for this condition. Adsorption granulocytapheresis may be an effective second-line therapy for Behcet, either as stand-alone treatment or in conjunction with other treatment modalities, but the supporting evidence consists only of observational studies or case series.(11) **(EG 2)**

For bullous pemphigoid and pemphigus vulgaris, For bullous pemphigoid, a systematic review concluded that adding plasma exchange to more standard treatment requires further investigation.(89) **(EG 1)** For pemphigus vulgaris, an expert consensus specialty society guideline states that the optimum roles of therapeutic plasma exchange, extracorporeal photophoresis, and immunoadsorption are not established for this condition.(11) **(EG 2)** Clinical studies of pemphigus vulgaris are largely limited to small series of patients in whom variable benefit has been observed after failure of more standard therapies. A single randomized controlled trial was not powered to answer the question of clinical benefit.(11)(90) **(EG 1)**

For burn shock resuscitation, An expert consensus specialty society guideline states that the optimum role of therapeutic plasma exchange is not established for this condition. Well-designed randomized controlled trials are needed to establish safety and efficacy; therapeutic plasma exchange for burn shock therapy is not recommended outside of clinical trials.(11) **(EG 2)** A review of therapeutic plasma exchange for burn shock concluded that while small studies suggest that this therapy may have beneficial effects in burn resuscitation, additional large prospective randomized studies comparing this therapy with conventional management are needed to establish the impact of therapeutic plasma exchange.(91) **(EG 2)**

For cardiac neonatal lupus, An expert consensus specialty society guideline states that the role of therapeutic plasma exchange is not established for this condition.(11) **(EG 2)**

For central retinal vein occlusion, A randomized controlled study of 61 patients that evaluated the efficacy of hemodilution using erythrocytapheresis found that, at 12-month follow-up, the erythrocytapheresis group was associated with improvement in visual acuity as well as reduction in conversion to the ischemic form.(92) **(EG 1)**

For coagulation factor inhibitors, An expert consensus specialty society guideline states that there is low-quality or very low-quality evidence supporting the use of therapeutic plasma exchange and immunoadsorption for this condition.(11) **(EG 2)**

For complex regional pain syndrome, An expert consensus specialty society guideline states that the role of therapeutic plasma exchange is not established for this condition.(11) **(EG 2)**

For dermatomyositis or polymyositis, An expert consensus specialty society guideline states that therapeutic plasma exchange and leukocytapheresis are ineffective for these conditions.(11) **(EG 2)** A systematic review evaluating treatments for dermatomyositis and polymyositis identified one study on the effects of plasmapheresis and leukapheresis and concluded that this intervention had no benefit in this setting.(93) **(EG 1)** No significant differences in final muscle strength or functional outcome were observed in patients with dermatomyositis or polymyositis randomly assigned to plasma exchange, leukapheresis, or sham treatment.(94) **(EG 2)**

For dilated cardiomyopathy (idiopathic), An expert consensus specialty society guideline states that the optimum role of therapeutic plasma exchange is not established for this condition. Small studies suggest that immunoadsorption improves clinical outcomes for patients with cardiac autoantibodies.(11) **(EG 2)** For post-transplant cardiomyopathy, a review article states that additional studies of therapeutic apheresis are needed to establish the frequency, duration, most appropriate technique, and type of replacement fluid needed for treatment.(95) **(EG 2)**

For encephalitis (chronic focal), An expert consensus specialty society guideline states that the optimum roles of therapeutic plasma exchange and immunoadsorption are not established for this condition.(11) **(EG 2)**

For encephalomyelitis (acute disseminated), An expert consensus specialty society guideline states that apheresis may be an effective therapy for acute disseminated encephalomyelitis, either as stand-alone treatment or in conjunction with other treatment modalities, but the supporting evidence consists only of case series and case reports.(11)(77) **(EG 2)**

For envenomation, An expert consensus specialty society guideline states that the optimum role of therapeutic plasma exchange is not established for this condition.(11) **(EG 2)**

For eosinophilic granulomatosis with polyangiitis, An expert consensus specialty society guideline states that the role of therapeutic plasma exchange has not been established for this condition.(11) **(EG 2)**

For erythropoietic porphyria with associated liver disease, An expert consensus specialty society guideline states that the roles of therapeutic plasma exchange and red blood cell exchange are not established for this condition.(11) **(EG 2)**

For Hashimoto encephalopathy (steroid-responsive encephalopathy associated with autoimmune thyroiditis), An expert consensus specialty society guideline states that therapeutic plasma exchange may be an effective second-line therapy for encephalopathy associated with autoimmune thyroiditis, either as a stand-alone treatment or in conjunction with other treatment modalities, but the supportive evidence consists only of observational studies and case reports.(11) **(EG 2)**

For hemolysis elevated liver enzymes and low platelets syndrome (HELLP syndrome), An expert consensus specialty society guideline states that the role of therapeutic plasma exchange is not established for this condition and may be harmful in the

antepartum period.(11) **(EG 2)** Because of pathophysiologic similarities to thrombotic thrombocytopenic purpura, plasmapheresis has been used in the management of HELLP syndrome; however, data for use in the antepartum period are limited to a few case series that showed mixed results and no clear benefit. A potential risk is fetal compromise due to reduction of already compromised placental blood flow.(96) **(EG 2)**

For hemolytic uremic syndrome, An expert consensus specialty society guideline states that therapeutic plasma exchange is not effective for adult or pediatric diarrhea-associated hemolytic uremic syndrome; the role of therapeutic plasma exchange or immunoadsorption has not been established in patients with associated severe neurologic symptoms. The use of plasma exchange as first-line therapy for atypical (not infection-associated) hemolytic uremic syndrome is based upon low-quality or very low-quality evidence.(11) **(EG 2)** Adults with hemolytic uremic syndrome have been treated with therapeutic plasma exchange due to high mortality and difficulty distinguishing between thrombotic thrombocytopenic purpura and hemolytic uremic syndrome.(53)(97) **(EG 2)**

For hemophagocytic lymphohistiocytosis, An expert consensus specialty society guideline states that the role of therapeutic plasma exchange has not been established for this condition.(11) **(EG 2)**

For Henoch-Schonlein purpura, An expert consensus specialty society guideline states that the optimum role of therapeutic plasma exchange is not established for this condition.(11) **(EG 2)**

For heparin-induced thrombocytopenia, An expert consensus specialty society guideline states that the optimum role of therapeutic plasma exchange is not established for this condition.(11) **(EG 2)**

For HLA desensitization prior to hematopoietic stem cell transplant, An expert consensus specialty society guideline states that the role of therapeutic plasma exchange is not established for this condition.(11) **(EG 2)**

For immune thrombocytopenia (refractory), An expert consensus specialty society guideline states that therapeutic plasma exchange is ineffective and that the optimum role of immunoadsorption is not established for this condition.(11) **(EG 2)**

For IgA nephropathy, An expert consensus specialty society guideline states that the optimum role of therapeutic plasma exchange is not established for this condition, including rapidly progressive crescentic glomerulonephritis, which develops in approximately 10% of cases. Some studies report that therapeutic plasma exchange appears to improve renal function during therapy and delay the time to dialysis dependence but does not arrest disease progression.(11) **(EG 2)**

For inclusion body myositis, An expert consensus specialty society guideline states that therapeutic plasma exchange and leukocytapheresis are ineffective for this condition. If a patient does not respond to steroids, intravenous immunoglobulin may have a transient effect, but treatment is generally symptomatic and supportive.(11) **(EG 2)**

For inflammatory bowel disease, An expert consensus specialty society guideline states that the optimum roles of adsorptive cytapheresis and extracorporeal photopheresis are not established for Crohn disease, and that adsorptive cytapheresis is not established for ulcerative colitis.(11) **(EG 2)** Several uncontrolled studies and reviews suggested that removal of activated circulating granulocytes and monocytes by apheresis may contribute to the amelioration of exacerbations of inflammatory bowel disease.(98)(99)(100)(101) **(EG 2)** A meta-analysis of 9 randomized controlled trials (686 patients) comparing standard pharmacotherapy with or without leukocytapheresis for active moderate to severe ulcerative colitis found that the combined regimen was associated with improved response, remission maintenance, and steroid-free patient rates. However, because of methodological problems with the majority of the studies, additional high-quality randomized controlled trials were recommended.(102) **(EG 1)** A meta-analysis of 9 randomized controlled trials (890 patients) found that granulocyte and monocyte adsorption apheresis was associated with increased clinical remission and response rates in patients with active, moderate, and severe ulcerative colitis as compared with treatment with corticosteroids. In addition, as compared to once-weekly apheresis, 2 or more sessions per week of apheresis was associated with an improved rate of clinical remission. Additional studies were

recommended to determine how and when to utilize granulocyte and monocyte adsorption apheresis as a therapeutic option for ulcerative colitis.(103) **(EG 1)** A systematic review concluded that more data are needed from trials with endoscopic evidence of active ulcerative colitis to define the role of apheresis in the management of this disease.(104) **(EG 1)** In a multicenter, randomized, double-blind, sham-controlled study of 235 adult patients with moderate to severe Crohn disease, granulocyte/monocyte apheresis did not demonstrate efficacy for induction of clinical remission or response.(105) **(EG 1)**

For Kawasaki disease, A study of 125 pediatric patients who were refractory to intravenous immunoglobulin and underwent plasma exchange found that the number of patients with coronary artery lesions (dilatation, aneurysm, or giant aneurysm) increased from 20 at baseline to 30 at 1 month and then decreased to 6 at 1 year. The authors stated that although the results were favorable, they were not statistically significant; in addition, the study was structurally limited because plasma exchange was not compared with other treatments such as steroids or infliximab, and the trial itself was not controlled.(106) **(EG 2)**

For Lambert-Eaton myasthenic syndrome, An expert consensus specialty society guideline states that there is low-quality or very low-quality evidence supporting the use of therapeutic plasma exchange for this condition, either as stand-alone treatment or in conjunction with other treatment modalities. No randomized controlled trials have demonstrated efficacy of therapeutic plasma exchange for this condition; however, several small uncontrolled case series have shown that some patients exhibit a transient improvement in symptoms.(11)(77) **(EG 2)**

For liver transplant, An observational study of 50 patients found that although plasma exchange significantly reduced total serum bilirubin and prothrombin time in patients with severe early liver transplant dysfunction, there were no significant reductions in liver enzymes and serum ammonia level. Graft survival at 6 months was 66%, with overall 1-year survival of 64%.(107) **(EG 2)**

For malaria, An expert consensus specialty society guideline states that the optimum role for red blood cell exchange is not established for severe malaria.(11) **(EG 2)** A review of the US National Malarial Surveillance System identified 101 patients who underwent exchange transfusion for the treatment of severe malaria and compared them to 314 matched patients who were treated without the intervention and found no difference in mortality between the groups. An associated literature review did not find evidence that exchange transfusion improved survival for patients with severe malaria.(108) **(EG 2)**

For multiple myeloma with renal failure at the time of diagnosis, A systematic review of 8 studies evaluating the efficacy of plasmapheresis for the treatment of myeloma kidney found no benefit of plasmapheresis independent of chemotherapy in terms of overall survival, recovery from dialysis, or improvement in renal function.(109) **(EG 1)** An expert consensus specialty society guideline states that therapeutic plasma exchange may be an effective second-line therapy for myeloma cast nephropathy, either as a stand-alone treatment or in conjunction with other treatment modalities, though in all cases survival depends on a satisfactory response to chemotherapy.(11) **(EG 2)** Expert opinion does not advocate the use of plasma exchange in such patients.(110)(111)(112) **(EG 2)**

For multiple sclerosis (chronic), An expert consensus specialty society guideline states that for chronic progressive multiple sclerosis, multiple randomized controlled trials demonstrate small to no benefit when therapeutic plasma exchange is used in conjunction with other immunosuppressive drugs; therapeutic plasma exchange has not been studied in relapsing-remitting multiple sclerosis.(11) **(EG 2)** Another expert consensus specialty society guideline states that therapeutic apheresis in primary and secondary chronic progressive disease is not effective.(6) **(EG 2)** Because of the lack of appropriate studies documenting efficacy, plasma exchange is not recommended as a permanent disease-modifying agent in multiple sclerosis patients.(29) **(EG 2)**

For natalizumab-associated progressive multifocal leukoencephalopathy, An expert consensus specialty society guideline states that there is low-quality or very low-quality evidence supporting therapeutic plasma exchange for natalizumab-associated progressive multifocal leukoencephalopathy, though it is acceptable first-line therapy, either as stand-alone treatment or in combination with other modalities. However, treatment with therapeutic plasma exchange is associated with immune reconstitution inflammatory syndrome.(11) **(EG 2)** A retrospective analysis of 219 patients with natalizumab-associated progressive multifocal

leukoencephalopathy who stopped natalizumab treatment found no difference in survival or clinical outcomes between patients who underwent plasma exchange (184 patients) compared with no further intervention.(113) **(EG 2)** Case reports also suggest that treatment of patients with natalizumab-associated progressive multifocal leukoencephalopathy with plasmapheresis and immunoadsorption may accelerate immune reconstitution inflammatory syndrome, with unclear effect on mortality.(114)(115) **(EG 2)**

For nephrogenic systemic fibrosis, An expert consensus specialty society guideline states that the optimum roles of extracorporeal photopheresis and therapeutic plasma exchange are not established for this condition.(11) **(EG 2)**

For pancreatitis due to hypertriglyceridemia, An expert consensus specialty society guideline states that the optimum role of therapeutic plasma exchange is not established for this condition. Although some small studies report that therapeutic apheresis can reduce hypertriglyceridemia and improve symptoms, a trial with historical controls (29 patients) found no difference between plasma exchange and standard medical therapy.(11) **(EG 2)** An observational study of 103 patients with 111 episodes of acute hypertriglyceridemic pancreatitis found that, as compared with conservative treatment, plasma exchange was associated with reduced levels of triglycerides. The authors noted that there are no controlled studies comparing conservative treatment and plasma exchange, and that although plasma exchange reduces triglycerides at a faster rate than conservative treatment, its impact on mortality remains unclear.(116) **(EG 2)**

For paraneoplastic neurologic syndromes, An expert consensus specialty society guideline states that the optimum roles of therapeutic plasma exchange and immunoadsorption are not established for this condition.(11) **(EG 2)**

For peripheral arterial disease, An expert consensus specialty society guideline states that LDL apheresis may be an acceptable second-line therapy for peripheral arterial disease, either alone or in combination with other treatment modalities.(11) **(EG 2)** A critical review found that there is insufficient evidence to support the use of lipid apheresis for patients with claudication or chronic limb ischemia.(117) **(EG 2)**

For polyneuropathy, organomegaly, endocrinopathy, M protein, and skin changes (POEMS), An expert consensus specialty society guideline states that therapeutic plasma exchange is ineffective for this condition.(11) **(EG 2)**

For post-transfusion purpura, An expert consensus specialty society guideline states that the optimum role of therapeutic plasma exchange is not established for this condition.(11) **(EG 2)**

For pruritus due to hepatobiliary disease, An expert consensus specialty society guideline states that the role of therapeutic plasma exchange has not been established for this condition.(11) **(EG 2)**

For psoriasis, An expert consensus specialty society guideline states that therapeutic plasma exchange is ineffective and that the optimum roles of adsorptive cytapheresis, lymphocytapheresis, and extracorporeal photopheresis are not established for this condition.(11) **(EG 2)**

For red cell alloimmunization during pregnancy, An expert consensus specialty society guideline states that the optimum role of therapeutic plasma exchange is not established for this condition.(11) **(EG 2)**

For red cell alloimmunization prevention after transfusion with incompatible blood, An expert consensus specialty society guideline states that the role of red blood cell exchange is not established for this condition.(11) **(EG 2)**

For rheumatoid arthritis, An expert consensus specialty society guideline states that published evidence suggests that apheresis is ineffective for this condition.(11) **(EG 2)** Noting that biological agents are currently the mainstay of rheumatoid arthritis treatment, a study of 4 patients with a reduced response to infliximab who underwent leukocytapheresis concluded that, although all 4 patients had a moderate response, more patients needed to be studied.(118) **(EG 2)** A randomized controlled trial of 60 patients (not on

steroids or biological agents) comparing plasmapheresis and disease-modifying antirheumatic drugs (DMARDs) to DMARDs alone found, at 6-month follow-up, that the combined therapy group was associated with increased response and remission rates as well as increased physical function. All patients in the combined therapy group achieved remission. The authors concluded that a multicenter randomized trial with longer follow-up was needed to confirm the findings.(119) **(EG 1)** A prospective study of 85 patients with rheumatoid arthritis resistant to treatment with DMARDs found that patients had significant improvement in the number of painful and swollen joints after 5 treatments of leukocytapheresis, and this improvement persisted for 4 weeks after therapy was completed. However, the authors cautioned that pretreatment heterogeneity and lack of randomization limit conclusions about efficacy and recommended further studies.(120) **(EG 2)**

For schizophrenia, An expert consensus specialty society guideline states that there are no studies addressing the efficacy of therapeutic plasma exchange for this condition.(11) **(EG 2)**

For scleroderma, An expert consensus specialty society guideline states that the optimum roles of therapeutic plasma exchange and extracorporeal photopheresis are not established for this condition.(11) **(EG 2)**

For sensorineural hearing loss (sudden), An expert consensus specialty society guideline states that the optimum roles of LDL apheresis, rheopheresis, and therapeutic plasma exchange are not established for this condition.(11) **(EG 2)** An observational study of 217 patients with sensorineural hearing loss who had not previously responded to steroids or plasma expanders found that, at a mean interval of 27 days between the onset of symptoms and a single session of fibrinogen/LDL apheresis, 15% had complete remission, 46% had partial remission, 33% had no change, and 2% worsened. It was also found that 32% of patients achieved full remission if apheresis occurred within 14 days of symptom onset, while none achieved full remission if apheresis occurred more than 90 days from symptom onset.(121) **(EG 2)**

For sepsis with multiorgan failure, A systematic review and meta-analysis of 4 studies (including 194 patients) found insufficient evidence to support the use of plasma exchange for the treatment of sepsis or septic shock.(122) **(EG 1)** An expert consensus specialty society guideline states that the optimum role of therapeutic plasma exchange is not established for this condition .(11) **(EG 2)**

For sickle cell disease (nonacute), pregnant patient, An expert consensus specialty society guideline states that the optimum role of red blood cell exchange is not established for this condition during pregnancy.(11) **(EG 2)**

For sickle cell disease (nonacute), preoperative management, An expert consensus specialty society guideline states that the optimum role of red blood cell exchange is not established for preoperative management of sickle cell disease.(11) **(EG 2)**

For sickle cell disease (nonacute), recurrent vaso-occlusive pain crisis, An expert consensus specialty society guideline states that the optimum role of red blood cell exchange is not established for this condition.(11) **(EG 2)**

For stiff-person syndrome, An expert consensus specialty society guideline states that the optimum role of therapeutic plasma exchange is not established for this condition. Evidence consists of only a few case studies with conflicting outcomes and no randomized trials.(11)(123)(124) **(EG 2)**

For Sydenham chorea, Expert consensus specialty society guidelines state that there is insufficient evidence to support the use of therapeutic plasma exchange for this condition.(11)(6) **(EG 2)**

For systemic lupus erythematosus, Despite early positive reports, randomized controlled trials have been unable to document a benefit of plasmapheresis as an adjunct to standard immunosuppressive therapy.(11)(31) **(EG 2)** A systematic review of 44 case series and reports evaluating therapies for diffuse alveolar hemorrhage in systemic lupus erythematosus found that plasmapheresis

did not influence survival.(125) **(EG 1)** A specialty society guideline states that there is inadequate evidence supporting the use of plasmapheresis for severe systemic lupus erythematosus.(126) **(EG 2)**

For thrombotic microangiopathy (coagulation mediated), An expert consensus specialty society guideline states that the role of therapeutic plasma exchange is not established for this condition.(11) **(EG 2)**

For thrombotic microangiopathy (refractory and associated with hematopoietic stem cell transplant), An expert consensus specialty society guideline states that the optimum role of therapeutic plasma exchange is not established for this condition.(11) **(EG 2)**

For thyroid storm, An expert consensus specialty society guideline states that the optimum role of therapeutic plasma exchange is not established for this condition.(11) **(EG 2)**

For toxic epidermal necrolysis (refractory), An expert consensus specialty society guideline states that the optimum role of therapeutic plasma exchange is not established for this condition.(11) **(EG 2)**

For transverse myelitis, A retrospective cohort study of 122 patients suggested, but did not confirm, that plasma exchange may provide some benefit.(127) **(EG 2)**

Reviewer Guidance

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For **Medicare** plans: Apply the current National Coverage Determination (NCD) for Extracorporeal Photopheresis 110.4 guidelines. See References section for full title and review dates.

Note: Prior authorization is not required for Photopheresis to treat Graft versus Host Disease (ICD D89.813). Because the CPT code is the same for the procedure, regardless of the diagnosis, the indication is included in the guideline. For claims processing purposes, do not void the case - no preauth. Rather, approve the request.

Note: 0342T is non-covered for any plan. Medicare: Carrier priced per Medicare Physician Fee Schedule (MPFS) - carrier makes determination and non-covered per UHC guideline MPG043.33. Not payable for RMHP PRIME (Medicaid) and CHP+ per HCPF guidance. Not a benefit / investigational for Individual and Family Plan (IFP) Commercial.

Policy History

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4/20/2023 Annual review and updates.

3/27/2022 Annual review and update to 25th edition MCG with NCD for Medicare and RMHP customizations.

Summary: 7/29/2020 RMHP adopted MCG guidelines with NCD for Medicare and retired the previous policy. 4/26/2021 Annual review – no changes.

References

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The Center for Medicare and Medicaid Services (CMS) National Coverage Determination (NCD) for Extracorporeal Photopheresis 110.4, effective date 4/30/2012, implementation date 10/1/2012, reviewed 4/10/2023.

Description

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The physician draws a patient's blood and exposes the blood to light to eliminate destructive elements. The physician establi shes venous access or attaches the machine to an existing central venous catheter line. The blood is removed and cycled through the pheresis machine where it is exposed to therapeutic wavelengths of light. The conditioned blood is returned to the patient th rough a catheter and a needle inserted in the vein.

References

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1. Padmanabhan A, et al. Guidelines on the use of therapeutic apheresis in clinical practice - evidence-based approach from the writing committee of the American Society for Apheresis: the eighth special issue. Journal of Clinical Apheresis 2019;34(3):171-354. DOI: 10.1002/jca.21705. [Context Link 1]
2. Daga Ruiz D, Fonseca San Miguel F, Gonzalez de Molina FJ, Ubeda-Iglesias A, Navas Perez A, Jannone Fores R. Plasmapheresis and other extracorporeal filtration techniques in critical patients. Medicina Intensiva 2017;41(3):174-187. DOI: 10.1016/j.medin.2016.10.005. [Context Link 1]
3. Sanchez AP, Cunard R, Ward DM. The selective therapeutic apheresis procedures. Journal of Clinical Apheresis 2013;28(1):20-29. DOI: 10.1002/jca.21265. [Context Link 1]
4. Winters JL. Randomized controlled trials in therapeutic apheresis. Journal of Clinical Apheresis 2013;28(1):48-55. DOI: 10.1002/jca.21263. [Context Link 1]
5. Howell C, et al. Guideline on the clinical use of apheresis procedures for the treatment of patients and collection of cellular therapy products. British Committee for Standards in Haematology. Transfusion Medicine 2015;25(2):57-78. DOI: 10.1111/tme.12205. (Reaffirmed 2020 Jun) [Context Link 1]
6. Cortese I, Chaudhry V, So YT, Cantor F, Cornblath DR, Rae-Grant A. Evidence-based guideline update: plasmapheresis in neurologic disorders: report of the Therapeutics and Technology Assessment Subcommittee of the American Academy of Neurology. Neurology 2011 (AAN reaffirmed 2015);76(3):294-300. DOI: 10.1212/WNL.0b013e318207b1f6. (Reaffirmed 2020 Nov) [Context Link 1, 2, 3, 4, 5]
7. Chevret S, Hughes RA, Annane D. Plasma exchange for Guillain-Barre syndrome. Cochrane Database of Systematic Reviews 2017, (verified by Cochrane 2017 Mar), Issue 2. Art. No.: CD001798. DOI: 10.1002/14651858.CD001798.pub3. [Context Link 1, 2]
8. Legault K, et al. McMaster RARE-Bestpractices clinical practice guideline on diagnosis and management of the catastrophic antiphospholipid syndrome. Journal of Thrombosis and Haemostasis 2018;Online. DOI: 10.1111/jth.14192. [Context Link 1]
9. Garcia D, Erkan D. Diagnosis and management of the antiphospholipid syndrome. New England Journal of Medicine 2018;378(21):2010-2021. DOI: 10.1056/NEJMra1705454. [Context Link 1]
10. Sanders JS, Rutgers A, Stegeman CA, Kallenberg CG. Pulmonary: renal syndrome with a focus on anti-GBM disease. Seminars in Respiratory and Critical Care Medicine 2011;32(3):328-334. DOI: 10.1055/s-0031-1279829. [Context Link 1, 2]
11. Casian A, Jayne D. Plasma exchange in the treatment of Wegener's granulomatosis, microscopic polyangiitis, Churg-Strauss syndrome and renal limited vasculitis. Current Opinion in Rheumatology 2011;23(1):12-17. DOI: 10.1097/BOR.0b013e32834120c1. [Context Link 1, 2]
12. Walsh M. Plasma exchange in antineutrophil cytoplasm antibody-associated vasculitis. Current Opinion in Nephrology and Hypertension 2014;23(6):555-559. DOI: 10.1097/MNH.0000000000000058. [Context Link 1]
13. Rosenfeld MR, Titulaer MJ, Dalmau J. Paraneoplastic syndromes and autoimmune encephalitis: Five new things. Neurology. Clinical Practice 2012;2(3):215-223. DOI: 10.1212/CPJ.0b013e31826af23e. [Context Link 1, 2]
14. Vannier E, Gelfand JA. Babesia species. In: Bennett JE, Dolin R, Blaser MJ, editors. Mandell, Douglas, and Bennett's Principles and Practice of Infectious Diseases. 9th ed. Philadelphia, PA: Elsevier; 2020:3400-3409.e2. [Context Link 1]

15. Mehndiratta MM, Hughes RA, Pritchard J. Plasma exchange for chronic inflammatory demyelinating polyradiculoneuropathy. Cochrane Database of Systematic Reviews 2015, (verified by Cochrane 2017 Nov), Issue 8. Art. No.: CD003906. DOI: 10.1002/14651858.CD003906.pub4. [Context Link 1, 2]

16. Gorson KC, Katz J. Chronic inflammatory demyelinating polyneuropathy. Neurologic Clinics 2013;31(2):511-532. DOI: 10.1016/j.ncl.2013.01.006. [Context Link 1]

17. Bunschoten C, Jacobs BC, Van den Bergh PYK, Cornblath DR, van Doorn PA. Progress in diagnosis and treatment of chronic inflammatory demyelinating polyradiculoneuropathy. Lancet Neurology 2019;18(8):784-794. DOI: 10.1016/S1474-4422(19)30144-9. [Context Link 1]

18. Mahr A, Chaigne-Delalande S, De Menthon M. Therapeutic plasma exchange in systemic vasculitis: an update on indications and results. Current Opinion in Rheumatology 2012;24(3):261-266. DOI: 10.1097/BOR.0b013e3283526509. [Context Link 1, 2]

19. Lee WP, Datta BN, Ong BB, Rees A, Halcox J. Defining the role of lipoprotein apheresis in the management of familial hypercholesterolemia. American Journal of Cardiovascular Drugs 2011;11(6):363-370. DOI: 10.2165/11594970-000000000-00000. [Context Link 1, 2]

20. Thompson GR, et al. Efficacy criteria and cholesterol targets for LDL apheresis. Atherosclerosis 2010;208(2):317-321. DOI: 10.1016/j.atherosclerosis.2009.06.010. [Context Link 1, 2]

21. Thompson GR. The evidence-base for the efficacy of lipoprotein apheresis in combating cardiovascular disease. Atherosclerosis. Supplements 2013;14(1):67-70. DOI: 10.1016/j.atherosclerosissup.2012.10.001. [Context Link 1]

22. Thompson GR, et al. Severe hypercholesterolaemia: therapeutic goals and eligibility criteria for LDL apheresis in Europe. Current Opinion in Lipidology 2010;21(6):492-498. DOI: 10.1097/MOL.0b013e3283402f53. [Context Link 1, 2, 3]

23. Page MM, Bell DA, Hooper AJ, Watts GF, Burnett JR. Lipoprotein apheresis and new therapies for severe familial hypercholesterolemia in adults and children. Best Practice & Research. Clinical Endocrinology & Metabolism 2014;28(3):387-403. DOI: 10.1016/j.beem.2013.10.004. [Context Link 1, 2, 3]

24. Palcoux JB, et al. Low-density lipoprotein apheresis in children with familial hypercholesterolemia: follow-up to 21 years. Therapeutic Apheresis and Dialysis 2008;12(3):195-201. DOI: 10.1111/j.1744-9987.2008.00574.x. [Context Link 1, 2]

25. Graedal A, et al. Apheresis in homozygous familial hypercholesterolemia: the results of a follow-up of all Norwegian patients with homozygous familial hypercholesterolemia. Journal of Clinical Lipidology 2012;6(4):331-339. DOI: 10.1016/j.jacl.2012.03.004. [Context Link 1]

26. Ganzel C, Becker J, Mintz PD, Lazarus HM, Rowe JM. Hyperleukocytosis, leukostasis and leukapheresis: practice management. Blood Reviews 2012;26(3):117-122. DOI: 10.1016/j.blre.2012.01.003. [Context Link 1]

27. Stone MJ, Bogen SA. Role of plasmapheresis in Waldenstrom's macroglobulinemia. Clinical Lymphoma, Myeloma & Leukemia 2013;13(2):238-240. DOI: 10.1016/j.clml.2013.02.013. [Context Link 1, 2, 3]

28. Tumani H. Corticosteroids and plasma exchange in multiple sclerosis. Journal of Neurology 2008;255 Suppl 6:36-42. DOI: 10.1007/s00415-008-6007-9. [Context Link 1, 2]

29. Gwathmey K, Balogun RA, Burns T. Neurologic indications for therapeutic plasma exchange: 2013 update. Journal of Clinical Apheresis 2014;29(4):211-219. DOI: 10.1002/jca.21331. [Context Link 1, 2, 3, 4, 5]

30. Balogun RA, et al. Clinical applications of therapeutic apheresis. Journal of Clinical Apheresis 2010;25(5):250-264. DOI: 10.1002/jca.20249. [Context Link 1, 2, 3, 4]

31. Ortiz-Salas P, Velez-Van-Meerbeke A, Galvis-Gomez CA, Rodriguez Q JH. Human immunoglobulin versus plasmapheresis in Guillain-Barre syndrome and myasthenia gravis: a meta-analysis. Journal of Clinical Neuromuscular Disease 2016;18(1):1-11. DOI: 10.1097/CND.0000000000000119. [Context Link 1, 2, 3]

32. Schroder A, Linker RA, Gold R. Plasmapheresis for neurological disorders. Expert Review of Neurotherapeutics 2009;9(9):1331-1339. DOI: 10.1586/ern.09.81. [Context Link 1]

33. Triantafyllou NI, Grapsa EI, Kararizou E, Psimenou E, Lagguranis A, Dimopoulos A. Periodic therapeutic plasma exchange in patients with moderate to severe chronic myasthenia gravis non-responsive to immunosuppressive agents: an eight year follow-up. Therapeutic Apheresis and Dialysis 2009;13(3):174-178. DOI: 10.1111/j.1744-9987.2009.00684.x. [Context Link 1, 2, 3]

34. Chaudhuri A, Behan PO. Myasthenic crisis. Quarterly Journal of Medicine 2009;102(2):97-107. DOI: 10.1093/qjmed/hcn152. [Context Link 1]

35. El-Bawab H, Hajjar W, Rafay M, Bamousa A, Khalil A, Al-Kattan K. Plasmapheresis before thymectomy in myasthenia gravis: routine versus selective protocols. European Journal of Cardio-Thoracic Surgery 2009;35(3):392-397. DOI: 10.1016/j.ejcts.2008.11.006. [Context Link 1, 2]

36. Keehn CA, Belongie IP, Shistik G, Fenske NA, Glass LF. The diagnosis, staging, and treatment options for mycosis fungoides. Cancer Control 2007;14(2):102-111. [Context Link 1]

37. Bosch X, Guilabert A, Espinosa G, Mirapeix E. Treatment of antineutrophil cytoplasmic antibody associated vasculitis: a systematic review. Journal of the American Medical Association 2007;298(6):655-669. DOI: 10.1001/jama.298.6.655. [Context Link 1]

38. Guillevin L, Pagnoux C. Indication for plasma exchange for systemic necrotizing vasculidities. Transfusion and Apheresis Science 2007;36(2):179-185. DOI: 10.1016/j.transci.2007.01.006. [Context Link 1, 2, 3, 4]

39. Guillevin L, Pagnoux C. Therapeutic strategies for systemic necrotizing vasculitides. Allergology International 2007;56(2):105-111. DOI: 10.2332/allergolint.R-07-144. [Context Link 1, 2]

40. Lehmann HC, Hartung HP, Meyer Zu Horste G, Kieseier BC. Plasma exchange in immune-mediated neuropathies. Current Opinion in Neurology 2008;21(5):547-554. DOI: 10.1097/WCO.0b013e32830b0f61. [Context Link 1, 2]

41. Carter CE, Benador NM. Therapeutic plasma exchange for the treatment of pediatric renal diseases in 2013. Pediatric Nephrology 2014;29(1):35-50. DOI: 10.1007/s00467-013-2479-7. [Context Link 1, 2, 3]

42. George SM, Balogun RA, Sanoff SL. Therapeutic apheresis before and after kidney transplantation. Journal of Clinical Apheresis 2011;26(5):252-260. DOI: 10.1002/jca.20297. [Context Link 1, 2]

43. Kahwaji J, Vo AA, Jordan SC. ABO blood group incompatibility: a diminishing barrier to successful kidney transplantation? Expert Review of Clinical Immunology 2010;6(6):893-900. DOI: 10.1586/eci.10.78. [Context Link 1]

44. Montgomery RA, et al. Desensitization in HLA-incompatible kidney recipients and survival. New England Journal of Medicine 2011;365(4):318-326. DOI: 10.1056/NEJMoa1012376. [Context Link 1, 2]

45. Nishio-Lucar A, Balogun RA, Sanoff S. Therapeutic apheresis in kidney transplantation: a review of renal transplant immunobiology and current interventions with apheresis medicine. Journal of Clinical Apheresis 2013;28(1):56-63. DOI: 10.1002/jca.21268. [Context Link 1, 2]

46. Coppo P, Veyradier A. Current management and therapeutical perspectives in thrombotic thrombocytopenic purpura. Presse Medicale 2012;41(3 Pt 2):e163-e176. DOI: 10.1016/j.lpm.2011.10.024. [Context Link 1, 2]

47. Nguyen TC, Han YY. Plasma exchange therapy for thrombotic microangiopathies. Organogenesis 2011;7(1):28-31. [Context Link 1, 2]

48. Frawley N, Ng AP, Nicholls K, Cohney S, Hogan C, Grigg A. Thrombotic thrombocytopenic purpura is associated with a high relapse rate after plasma exchange: a single-centre experience. Internal Medicine Journal 2009;39(1):19-24. DOI: 10.1111/j.1445-5994.2008.01637.x. [Context Link 1, 2]

49. Okafor C, Ward DM, Mokrzycki MH, Weinstein R, Clark P, Balogun RA. Introduction and overview of therapeutic apheresis. Journal of Clinical Apheresis 2010;25(5):240-249. DOI: 10.1002/jca.20247. [Context Link 1, 2]

50. Reeves HM, Winters JL. The mechanisms of action of plasma exchange. British Journal of Haematology 2014;164(3):342-351. DOI: 10.1111/bjh.12629. [Context Link 1, 2]

51. Dierickx D, Macken E. The ABC of apheresis. Acta Clinica Belgica 2015;70(2):95-99. DOI: 10.1179/2295333714Y.0000000096. [Context Link 1, 2]

52. Kaplan AA. Therapeutic plasma exchange: core curriculum 2008. American Journal of Kidney Diseases 2008;52(6):1180-1196. DOI: 10.1053/j.ajkd.2008.02.360. [Context Link 1, 2, 3, 4]

53. Rodriguez-Pinto I, Espinosa G, Erkan D, Shoenfeld Y, Cervera R, CAPS Registry Project Group. The effect of triple therapy on the mortality of catastrophic anti-phospholipid syndrome patients. Rheumatology 2018;57(7):1264-1270. DOI: 10.1093/rheumatology/key082. [Context Link 1]

54. Blaha M, et al. The importance of rheological parameters in the therapy of the dry form of age-related macular degeneration with rheohaemapheresis. Clinical Hemorheology and Microcirculation 2012;50(4):245-255. DOI: 10.3233/CH-2011-1431. [Context Link 1]

55. Baweja S, Wiggins K, Lee D, Blair S, Fraenkel M, McMahon LP. Benefits and limitations of plasmapheresis in renal diseases: an evidence-based approach. Journal of Artificial Organs 2011;14(1):9-22. DOI: 10.1007/s10047-010-0529-5. [Context Link 1]

56. Silvarino R, Noboa O, Cervera R. Anti-glomerular basement membrane antibodies. Israel Medical Association Journal 2014;16(11):727-732. [Context Link 1]

57. Balogun RA, et al. Update to the ASFA guidelines on the use of therapeutic apheresis in ANCA-associated vasculitis. Journal of Clinical Apheresis 2020;Online. DOI: 10.1002/jca.21820. [Context Link 1]

58. Derebail VK, Falk RJ. ANCA-associated vasculitis - refining therapy with plasma exchange and glucocorticoids. New England Journal of Medicine 2020;382(7):671-673. DOI: 10.1056/NEJMe1917490. [Context Link 1]

59. Walters GD, Willis NS, Cooper TE, Craig JC. Interventions for renal vasculitis in adults. Cochrane Database of Systematic Reviews 2020, Issue 1. Art. No.: CD003232. DOI: 10.1002/14651858.CD003232.pub4. [Context Link 1]

60. Walsh M, et al. Long-term follow-up of patients with severe ANCA-associated vasculitis comparing plasma exchange to intravenous methylprednisolone treatment is unclear. Kidney International 2013;84(2):397-402. DOI: 10.1038/ki.2013.131. [Context Link 1]

61. Suppiej A, et al. Plasma exchange in pediatric anti-NMDAR encephalitis: A systematic review. Brain and Development 2016;38(7):613-622. DOI: 10.1016/j.braindev.2016.01.009. [Context Link 1]

62. Zuliani L, Graus F, Giometto B, Bien C, Vincent A. Central nervous system neuronal surface antibody associated syndromes: review and guidelines for recognition. Journal of Neurology, Neurosurgery, and Psychiatry 2012;83(6):638-645. DOI: 10.1136/jnnp-2011-301237. [Context Link 1]

63. Colvin MM, et al. Sensitization in heart transplantation: emerging knowledge: a scientific statement from the American Heart Association. Circulation 2019;139(12):e553-e578. DOI: 10.1161/CIR.0000000000000598. [Context Link 1]

64. McMillan HJ, Kang PB, Jones HR, Darras BT. Childhood chronic inflammatory demyelinating polyradiculoneuropathy: combined analysis of a large cohort and eleven published series. Neuromuscular Disorders: NMD 2013;23(2):103-111. DOI: 10.1016/j.nmd.2012.09.008. [Context Link 1]

65. Dammacco F, Sansonno D. Therapy for hepatitis C virus-related cryoglobulinemic vasculitis. New England Journal of Medicine 2013;369(11):1035-1045. DOI: 10.1056/NEJMra1208642. [Context Link 1]

66. Montero N, et al. Treatment for hepatitis C virus-associated mixed cryoglobulinaemia. Cochrane Database of Systematic Reviews 2018, Issue 5. Art. No.: CD011403. DOI: 10.1002/14651858.CD011403.pub2. [Context Link 1]

67. Brederson C, Rumble RB, Varela NP, Kuruvilla J, Kouroukis CT, Stem Cell Transplant Steering Committee. Extra-Corporeal Photopheresis in the Management of Graft-Versus-Host Disease in Patients Who Have Received Allogeneic Blood or Bone Marrow Transplants: Recommendations. [Internet] Cancer Care Ontario. 2013 Aug Accessed at: https://www.cancercareontario.ca/. [accessed 2020 Aug 19] [Context Link 1]

68. Buzzetti E, Kalafateli M, Thorburn D, Davidson BR, Tsochatzis E, Gurusamy KS. Interventions for hereditary haemochromatosis. Cochrane Database of Systematic Reviews 2017, Issue 3. Art. No.: CD011647. DOI: 10.1002/14651858.CD011647.pub2. [Context Link 1]

69. Winters JL. Lipid apheresis, indications, and principles. Journal of Clinical Apheresis 2011;26(5):269-275. DOI: 10.1002/jca.20299. [Context Link 1]

70. France M, et al. HEART UK statement on the management of homozygous familial hypercholesterolaemia in the United Kingdom. Atherosclerosis 2016;255:128-139. DOI: 10.1016/j.atherosclerosis.2016.10.017. [Context Link 1]

71. Kuo KH, et al. A retrospective observational study of leucoreductive strategies to manage patients with acute myeloid leukaemia presenting with hyperleucocytosis. British Journal of Haematology 2015;168(3):384-394. DOI: 10.1111/bjh.13146. [Context Link 1]

72. Oberoi S, et al. Leukapheresis and low-dose chemotherapy do not reduce early mortality in acute myeloid leukemia hyperleukocytosis: a systematic review and meta-analysis. Leukemia Research 2014;38(4):460-468. DOI: 10.1016/j.leukres.2014.01.004. [Context Link 1]

73. Owen RG, et al. Guidelines on the diagnosis and management of Waldenstrom macroglobulinaemia. British Journal of Haematology 2014;165(3):316-333. DOI: 10.1111/bjh.12760. (Reaffirmed 2020 Jun) [Context Link 1]

74. Khan TZ, et al. Apheresis as novel treatment for refractory angina with raised lipoprotein(a): a randomized controlled cross-over trial. European Heart Journal 2017;38(20):1561-1569. DOI: 10.1093/eurheartj/ehx178. [Context Link 1]

75. Larsen FS, et al. High-volume plasma exchange in patients with acute liver failure: An open randomised controlled trial. Journal of Hepatology 2016;64(1):69-78. DOI: 10.1016/j.jhep.2015.08.018. [Context Link 1]

76. Cortese I, Cornblath DR. Therapeutic plasma exchange in neurology: 2012. Journal of Clinical Apheresis 2013;28(1):16-19. DOI: 10.1002/jca.21266. [Context Link 1, 2, 3, 4, 5, 6, 7, 8]

77. Mandawat A, Mandawat A, Kaminski HJ, Shaker ZA, Alawi AA, Alshekhlee A. Outcome of plasmapheresis in myasthenia gravis: delayed therapy is not favorable. Muscle and Nerve 2011;43(4):578-584. DOI: 10.1002/mus.21924. [Context Link 1]

78. Horwitz SM, et al. Primary Cutaneous Lymphomas. NCCN Clinical Practice Guidelines in Oncology [Internet] National Comprehensive Cancer Network (NCCN). v. 1.2021; 2020 Oct Accessed at: https://www.nccn.org/professionals/physician_gls/f_guidelines.asp. [accessed 2020 Oct 14] [Context Link 1]

79. Chou ST, et al. American Society of Hematology 2020 guidelines for sickle cell disease: transfusion support. Blood Advances 2020;4(2):327-355. DOI: 10.1182/bloodadvances.2019001143. [Context Link 1]

80. DeBaun MR, et al. American Society of Hematology 2020 guidelines for sickle cell disease: prevention, diagnosis, and treatment of cerebrovascular disease in children and adults. Blood Advances 2020;4(8):1554-1588. DOI: 10.1182/bloodadvances.2019001142. [Context Link 1]

81. DeBaun MR, et al. Controlled trial of transfusions for silent cerebral infarcts in sickle cell anemia. New England Journal of Medicine 2014;371(8):699-710. DOI: 10.1056/NEJMoa1401731. [Context Link 1]

82. Adams RJ, et al. Prevention of a first stroke by transfusions in children with sickle cell anemia and abnormal results on transcranial Doppler ultrasonography. New England Journal of Medicine 1998;339(1):5-11. [Context Link 1]

83. Bavle A, Raj A, Kong M, Bertolone S. Impact of long-term erythrocytapheresis on growth and peak height velocity of children with sickle cell disease. Pediatric Blood and Cancer 2014;61(11):2024-2030. DOI: 10.1002/pbc.25153. [Context Link 1]

84. Squires RH, et al. Evaluation of the pediatric patient for liver transplantation: 2014 practice guideline by the American Association for the Study of Liver Diseases, American Society of Transplantation and the North American Society for Pediatric Gastroenterology, Hepatology and Nutrition. Hepatology 2014;60(1):362-398. DOI: 10.1002/hep.27191. (Reaffirmed 2020 Jun) [Context Link 1]

85. Sheppard D, et al. Major ABO-incompatible BMT: isohemagglutinin reduction with plasma exchange is safe and avoids graft manipulation. Bone Marrow Transplantation 2013;48(7):953-957. DOI: 10.1038/bmt.2012.264. [Context Link 1]

86. Stussi G, et al. Prevention of pure red cell aplasia after major or bidirectional ABO blood group incompatible hematopoietic stem cell transplantation by pretransplant reduction of host anti-donor isoagglutinins. Haematologica 2009;94(2):239-248. DOI: 10.3324/haematol.13356. [Context Link 1]

87. Lehmann HC, Hartung HP, Hetzel GR, Stuve O, Kieseier BC. Plasma exchange in neuroimmunological disorders. Part 2: treatment of neuromuscular disorders. Archives of Neurology 2006;63(8):1066-1071. DOI: 10.1001/archneur.63.8.1066. [Context Link 1]

88. Kirtschig G, Middleton P, Bennett C, Murrell DF, Wojnarowska F, Khumalo NP. Interventions for bullous pemphigoid. Cochrane Database of Systematic Reviews 2010, (verified by Cochrane 2016 Feb), Issue 10. Art. No.: CD002292. DOI: 10.1002/14651858.CD002292.pub3. [Context Link 1]

89. Sagi L, Baum S, Gendelman V, Trau H, Barzilai A. The role of therapeutic plasma exchange in pemphigus vulgaris. Journal of the European Academy of Dermatology and Venereology: JEADV. 2011;25(1):82-86. DOI: 10.1111/j.1468-3083.2010.03703.x. [Context Link 1]

90. Mosier MJ, DeChristopher PJ, Gamelli RL. Use of therapeutic plasma exchange in the burn unit: a review of the literature. Journal of Burn Care and Research 2013;34(3):289-298. DOI: 10.1097/BCR.0b013e318283d18c. [Context Link 1]

91. Glacet-Bernard A, et al. Hemodilution therapy using automated erythrocytapheresis in central retinal vein occlusion: results of a multicenter randomized controlled study. Graefe's Archive for Clinical and Experimental Ophthalmology 2011;249(4):505-512. DOI: 10.1007/s00417-010-1532-5. [Context Link 1]

92. Vermaak E, Tansley SL, McHugh NJ. The evidence for immunotherapy in dermatomyositis and polymyositis: a systematic review. Clinical Rheumatology 2015;34(12):2089-2095. DOI: 10.1007/s10067-015-3059-y. [Context Link 1]

93. Linker RA, Gold R. Use of intravenous immunoglobulin and plasma exchange in neurological disease. Current Opinion in Neurology 2008;21(3):358-365. DOI: 10.1097/WCO.0b013e3282ff5b8f. [Context Link 1]

94. Pignalosa O, Infante T, Napoli C. The use of therapeutic apheresis in cardiovascular disease. Transfusion Medicine 2014;24(2):68-78. DOI: 10.1111/tme.12103. [Context Link 1]

95. Maynard SE, Karumanchi SA, Thadhani RI. Pregnancy and kidney disease. In: Yu AS, Chertow GM, Luyckx VA, Marsden PA, Skorecki K, Taal MW, editors. Brenner and Rector's The Kidney. 11th ed. Philadelphia, PA: Elsevier; 2020:1622-1653.e12. [Context Link 1]

96. Forzley BR, Clark WF. TTP/HUS and prognosis: the syndrome and the disease(s). Kidney International. Supplement 2009;(112):S59-561. DOI: 10.1038/ki.2008.623. [Context Link 1]

97. Passalacqua S, et al. The Italian Registry of Therapeutic Apheresis: granulocyte-monocyte apheresis in the treatment of inflammatory bowel disease. A multicentric study. Journal of Clinical Apheresis 2011;26(6):332-337. DOI: 10.1002/jca.20315. [Context Link 1]

98. Ruuska T, et al. Granulocyte-monocyte adsorptive apheresis in pediatric inflammatory bowel disease: results, practical issues, safety, and future perspectives. Inflammatory Bowel Diseases 2009;15(7):1049-1054. DOI: 10.1002/ibd.20859. [Context Link 1]

99. Tomomasa T, et al. Leukocytapheresis in pediatric patients with ulcerative colitis. Journal of Pediatric Gastroenterology and Nutrition 2011;53(1):34-39. DOI: 10.1097/MPG.0b013e31821058bc. [Context Link 1]

100. Vernia P, D'Ovidio V, Meo D. Leukocytapheresis in the treatment of inflammatory bowel disease: Current position and perspectives. Transfusion and Apheresis Science 2010;43(2):227-229. DOI: 10.1016/j.transci.2010.07.023. [Context Link 1]

101. Zhu M, Xu X, Nie F, Tong J, Xiao S, Ran Z. The efficacy and safety of selective leukocytapheresis in the treatment of ulcerative colitis: a meta-analysis. International Journal of Colorectal Disease 2011;26(8):999-1007. DOI: 10.1007/s00384-011-1193-9. [Context Link 1]

102. Yoshino T, et al. Efficacy and safety of granulocyte and monocyte adsorption apheresis for ulcerative colitis: a meta-analysis. Digestive and Liver Disease 2014;46(3):219-226. DOI: 10.1016/j.dld.2013.10.011. [Context Link 1]

103. Thanaraj S, Hamlin PJ, Ford AC. Systematic review: granulocyte/monocyte adsorptive apheresis for ulcerative colitis. Alimentary Pharmacology and Therapeutics 2010;32(11-12):1297-1306. DOI: 10.1111/j.1365-2036.2010.04490.x. [Context Link 1]

104. Sands BE, et al. A randomised, double-blind, sham-controlled study of granulocyte/monocyte apheresis for moderate to severe Crohn's disease. Gut 2013;62(9):1288-1294. DOI: 10.1136/gutjnl-2011-300995. [Context Link 1]

105. Hokosaki T, et al. Long-term efficacy of plasma exchange treatment for refractory Kawasaki disease. Pediatrics International 2012;54(1):99-103. DOI: 10.1111/j.1442-200X.2011.03487.x. [Context Link 1]

106. Park CS, et al. Role of plasmapheresis as liver support for early graft dysfunction following adult living donor liver transplantation. Transplantation Proceedings 2012;44(3):749-751. DOI: 10.1016/j.transproceed.2012.01.054. [Context Link 1]

107. Tan KR, Wiegand RE, Arguin PM. Exchange transfusion for severe malaria: evidence base and literature review. Clinical Infectious Diseases 2013;57(7):923-928. DOI: 10.1093/cid/cit429. [Context Link 1]

108. Gupta D, Bachegowda L, Phadke G, Boren S, Johnson D, Misra M. Role of plasmapheresis in the management of myeloma kidney: a systematic review. Hemodialysis International 2010;14(4):355-363. DOI: 10.1111/j.1542-4758.2010.00481.x. [Context Link 1]

109. Cockwell P, Cook M. The rationale and evidence base for the direct removal of serum-free light chains in the management of myeloma kidney. Advances in Chronic Kidney Disease 2012;19(5):324-332. DOI: 10.1053/j.ackd.2012.06.003. [Context Link 1]

110. Movilli E, Guido J, Silvia T, Francesco S, Giovanni C. Plasma exchange in the treatment of acute renal failure of myeloma. Nephrology, Dialysis, Transplantation 2007;22(4):1270-1271. DOI: 10.1093/ndt/gfl628. [Context Link 1]

111. Cserti C, Haspel R, Stowell C, Dzik W. Light-chain removal by plasmapheresis in myeloma-associated renal failure. Transfusion 2007;47(3):511-514. DOI: 10.1111/j.1537-2995.2006.01143.x. [Context Link 1]

112. Landi D, et al. No evidence of beneficial effects of plasmapheresis in natalizumab-associated PML. Neurology 2017;88(12):1144-1152. DOI: 10.1212/WNL.0000000000003740. [Context Link 1]

113. Tan IL, McArthur JC, Clifford DB, Major EO, Nath A. Immune reconstitution inflammatory syndrome in natalizumab-associated PML. Neurology 2011;77(11):1061-1067. DOI: 10.1212/WNL.0b013e31822e55e7. [Context Link 1]

114. Clifford DB, et al. Natalizumab-associated progressive multifocal leukoencephalopathy in patients with multiple sclerosis: lessons from 28 cases. Lancet Neurology 2010;9(4):438-446. DOI: 10.1016/S1474-4422(10)70028-4. [Context Link 1]

115. Gubensek J, Buturovic-Ponikvar J, Romozi K, Ponikvar R. Factors affecting outcome in acute hypertriglyceridemic pancreatitis treated with plasma exchange: an observational cohort study. PLoS ONE 2014;9(7):e102748. DOI: 10.1371/journal.pone.0102748. [Context Link 1]

116. Weiss N. A critical review on the use of lipid apheresis and rheopheresis for treatment of peripheral arterial disease and the diabetic foot syndrome. Seminars in Dialysis 2012;25(2):220-227. DOI: 10.1111/j.1525-139X.2011.01036.x. [Context Link 1]

117. Sakai Y, et al. Efficacy of high-throughput leukocytapheresis for rheumatoid arthritis with a reduced response to infliximab. Therapeutic Apheresis and Dialysis 2009;13(3):179-185. DOI: 10.1111/j.1744-9987.2009.00657.x. [Context Link 1]

118. Yu X, et al. Effects of double filtration plasmapheresis, leflunomide, and methotrexate on inflammatory changes found through magnetic resonance imaging in early rheumatoid arthritis. Journal of Rheumatology 2012;39(6):1171-1178. DOI: 10.3899/jrheum.110978. [Context Link 1]

119. Kitagaichi M, et al. Safety and efficacy of the leukocytapheresis procedure in eighty-five patients with rheumatoid arthritis. Transfusion and Apheresis Science 2016;55(2):225-232. DOI: 10.1016/j.transci.2016.07.019. [Context Link 1]

120. Canis M, Heigl F, Suckfuell M. Fibrinogen/LDL apheresis is a promising rescue therapy for sudden sensorineural hearing loss. Clinical Research in Cardiology Supplements 2012;7(Suppl 1):36-40. DOI: 10.1007/s11789-012-0044-8. [Context Link 1]

121. Rimmer E, et al. The efficacy and safety of plasma exchange in patients with sepsis and septic shock: a systematic review and meta-analysis. Critical Care 2014;18(6):699. DOI: 10.1186/s13054-014-0699-2. [Context Link 1]

122. Pagano MB, Murinson BB, Tobian AA, King KE. Efficacy of therapeutic plasma exchange for treatment of stiff-person syndrome. Transfusion 2014;54(7):1851-1856. DOI: 10.1111/trf.12573. [Context Link 1]

123. Albahra S, Yates SG, Joseph D, De Simone N, Burner JD, Sarode R. Role of plasma exchange in stiff person syndrome. Transfusion and Apheresis Science 2019;58(3):310-312. DOI: 10.1016/j.transci.2019.03.015. [Context Link 1]

124. Ednalino C, Yip J, Carsons SE. Systematic review of diffuse alveolar hemorrhage in systemic lupus erythematosus: focus on outcome and therapy. Journal of Clinical Rheumatology 2015;21(6):305-310. DOI: 10.1097/RHU.0000000000000291. [Context Link 1]

125. Gordon C, et al. The British Society for Rheumatology guideline for the management of systemic lupus erythematosus in adults. Rheumatology 2018;57(1):e1-e45. DOI: 10.1093/rheumatology/kex286. (Reaffirmed 2020 Jun) [Context Link 1]

126. Greenberg BM, Thomas KP, Krishnan C, Kaplin AI, Calabresi PA, Kerr DA. Idiopathic transverse myelitis: corticosteroids, plasma exchange, or cyclophosphamide. Neurology 2007;68(19):1614-1617. DOI: 10.1212/01.wnl.0000260970.63493.c8. [Context Link 1]

Footnotes

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[A] Patients who present with a creatinine level less than 6.6 mg/dL (583 micromoles/L) can recover renal function after plasma exchange. Patients with a creatinine level greater than 6.6 mg/dL (583 micromoles/L) or who are dialysis dependent will not recover renal function after plasma exchange.(11) [A in Context Link 1]

[B] Babesiosis is a tick-borne infectious disease caused by an intraerythrocytic protozoan.(11) [B in Context Link 1, 2]

[C] A serum viscosity of greater than 4.0 (relative to water) is associated with an increased risk of hyperviscosity-related events.(28) [C in Context Link 1, 2]

[D] Photopheresis consists of the addition of psoralen to a bag of lymphocytes taken from the patient via pheresis. The psoralen-exposed cells are then exposed to UVA light and returned to the patient.(31)(37) [D in Context Link 1]

Codes

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