Plasmapheresis

Selecting a Treatment Regimen
Plasmapheresis, also known as therapeutic plasma exchange, is a procedure that involves separating the blood, exchanging the plasma (typically with donor plasma or albumin solution), and returning the other components, primarily red blood cells, to the patient.

The mechanics of plasmapheresis have not changed since the introduction of continuous flow machines.

### Selecting a Treatment Regimen

#### Acute Inflammatory Demyelinating Polyneuropathy (AIDP)/Guillain-Barré Syndrome (GBS)

- Plasmapheresis should be offered in the treatment of AIDP/GBS severe enough to impair independent walking or to require mechanical ventilation (Level A).

- Plasmapheresis should be considered in the treatment of milder clinical presentations of AIDP/GBS (Level B).
  
  Remark: IV immunoglobulin (IVIg) is an alternative treatment used in patients with AIDP/GBS. There is insufficient evidence to demonstrate the superiority of one treatment over the other.

#### Chronic Inflammatory Demyelinating Neuropathy (CIDP)

- Plasmapheresis should be offered as a short-term treatment for patients with CIDP (Level A).

  Remark: Steroids, IVIg, and immunosuppressants have also been used in the treatment of CIDP.

#### CNS Demyelinating Disease

- Based on a single, small Class II study, plasmapheresis is possibly effective for acute fulminant CNS demyelinating diseases (multiple sclerosis [MS], transverse myelitis [TM], acute disseminated encephalomyelitis [ADEM], Marburg variant, neuromyelitis optica [NMO], recurrent myelitis, focal cerebral demyelination) that fail to respond to high-dose corticosteroid treatment.

  Note: Because the study included subgroups of patients with demyelinating diseases, it is not possible to determine if plasmapheresis is more or less effective in patients with different demyelinating diseases.

- Plasmapheresis may be considered in the treatment of fulminant CNS demyelinating diseases that fail to respond to high-dose corticosteroid treatment (Level C).

### Multiple Sclerosis (MS)

Plasmapheresis as adjunctive therapy is probably effective for management of exacerbations in relapsing forms of MS, based on a single Class I study.

- Plasmapheresis should be considered for the adjunctive treatment of exacerbations in relapsing forms of MS (Level B).

- For chronic progressive or secondary progressive MS, plasmapheresis is established as ineffective based on consistent Class I evidence.

  Note: The term chronic progressive MS is no longer used, but previously included patients are now described as having either primary progressive MS or secondary progressive MS.

- Plasmapheresis should not be offered for chronic progressive or secondary progressive MS (Level A).

  Remark: No studies on the efficacy of plasmapheresis compared to other treatment options in MS are available.

### Dysimmune Neuropathies

One Class I study showed the efficacy of plasmapheresis in polyneuropathies associated with immunoglobulin A (IgA) and immunoglobulin G (IgG) monoclonal gammopathy of undetermined significance (MGUS). That study, and a Class III study, found no significant benefit in immunoglobulin M (IgM)-associated MGUS.

- Plasmapheresis should be considered in polyneuropathy associated with IgA and IgG MGUS (Level B).

- Plasmapheresis should **NOT** be considered in the treatment of polyneuropathy associated with IgM MGUS (Level B).

### Myasthenia Gravis (MG)

- Because of the lack of randomized controlled studies with masked outcomes, there is insufficient evidence to support or refute the efficacy of plasmapheresis in the treatment of myasthenic crisis (Level U) or MG prethymectomy (Level U).

  Remark: Despite the fact that the use of plasmapheresis in myasthenic crisis and MG prethymectomy receives a Level U recommendation, plasmapheresis is used at many medical centers for these indications.
Pediatric Autoimmune Neuropsychiatric Disorders Associated with Streptococcal Infection (PANDAS)

PANDAS is defined as the abrupt onset or exacerbation of a tic or obsessive-compulsive disorder (OCD) in prepubertal children, considered to be triggered by a Group A β-hemolytic streptococcal infection, but there is controversy in the medical community regarding this syndrome as a disease entity.

There is insufficient evidence to support or refute the use of plasmapheresis in the treatment of acute OCD and tic symptoms in the setting of PANDAS (Level U).

Sydenham Chorea

There is insufficient evidence to support or refute the use of plasmapheresis in the treatment of Sydenham chorea (Level U).

Table 1. Summary of Evidence

<table>
<thead>
<tr>
<th>Disease</th>
<th>Conclusion</th>
<th>Quality</th>
</tr>
</thead>
<tbody>
<tr>
<td>Acute Inflammatory Demyelinating Polynuropathy/Guillain-Barré Syndrome</td>
<td>Established effective</td>
<td>Class I</td>
</tr>
<tr>
<td>Chronic Inflammatory Demyelinating Polynuropathy, short-term treatment</td>
<td>Established effective</td>
<td>Class I</td>
</tr>
<tr>
<td>Polyneuropathy with monoclonal gammopathies of undetermined significance</td>
<td>IgA/IgG: Probably effective</td>
<td>Class I</td>
</tr>
<tr>
<td></td>
<td>IgM: Probably ineffective</td>
<td>Class I</td>
</tr>
<tr>
<td>Myasthenia Gravis</td>
<td>Pre-op Preparation: Insufficient evidence</td>
<td>Class III</td>
</tr>
<tr>
<td></td>
<td>Crisis: Insufficient evidence</td>
<td>Class III</td>
</tr>
<tr>
<td>Fulminant Demyelinating CNS Disease</td>
<td>Possibly effective</td>
<td>Class II</td>
</tr>
<tr>
<td>Chronic Progressive MS</td>
<td>Established ineffective</td>
<td>Class I</td>
</tr>
<tr>
<td>Relapses in MS</td>
<td>Probably effective</td>
<td>Class I</td>
</tr>
<tr>
<td>Sydenham Chorea</td>
<td>Insufficient evidence</td>
<td>Class III</td>
</tr>
<tr>
<td>Acute OCD and Tics in PANDAS</td>
<td>Insufficient evidence</td>
<td>Class III</td>
</tr>
</tbody>
</table>

Classification of Management Recommendations

<table>
<thead>
<tr>
<th>Classification</th>
<th>Definition</th>
</tr>
</thead>
<tbody>
<tr>
<td>Strong (A)</td>
<td>Established as effective, ineffective, or harmful (or established as useful/predictive or not useful/predictive) for the given condition in the specified population. (Level A rating requires at least two consistent Class I studies).*</td>
</tr>
<tr>
<td>Moderate (B)</td>
<td>Probably effective, ineffective, or harmful (or probably useful/predictive or not useful/predictive) for the given condition in the specified population. (Level B rating requires at least one Class I study or two consistent Class II studies.)</td>
</tr>
<tr>
<td>Weak (C)</td>
<td>Possibly effective, ineffective, or harmful (or possibly useful/predictive or not useful/predictive) for the given condition in the specified population. (Level C rating requires at least one Class II study or two consistent Class III studies.)</td>
</tr>
<tr>
<td>Insufficient (U)</td>
<td>Data inadequate or conflicting; given current knowledge, treatment (test, predictor) is unproven.</td>
</tr>
</tbody>
</table>

Note: In exceptional cases, one convincing Class I study may suffice for an “A” recommendation if 1) all criteria are met, 2) the magnitude of effect is large (relative rate improved outcome is greater than 5 and the lower limit of the confidence interval is greater than 2).

Source

Endorsed by the American Association of Neuromuscular and Electrodiagnostic Medicine. The National Multiple Sclerosis Society endorsed only the section that relates to the use of plasma exchange for multiple sclerosis.

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