

THERAPEUTIC PLASMA EXCHANGE

(TPE, Plasmapheresis, or PLEX)



Answers to questions you may have
about therapeutic plasma exchange

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Myasthenia gravis (MG) is an autoimmune disease. Under normal circumstances, the body's immune system protects against infection from invading bacteria and viruses. In autoimmune diseases like MG, the immune system produces antibodies that target normal tissues and cause undesirable effects. In MG, these antibodies currently include antiacetylcholine receptor (AChR) antibodies, anti-muscle-specific tyrosine kinase (MuSK) antibodies, low-density lipoprotein related protein 4 (LRP4) antibodies and others being researched. These antibodies attack proteins at the neuromuscular junction resulting in muscle weakness. During therapeutic plasma exchange, these abnormal antibodies are removed from the bloodstream, leading to improvement in muscle strength.

What is therapeutic plasma exchange?

Therapeutic plasma exchange is a treatment for antibody-mediated diseases like MG. During this procedure, blood is separated into cells and plasma (liquid). The plasma portion is removed and typically replaced with either a human-derived sterile fluid called albumin or plasma. The procedure is often referred to as plasma exchange (PLEX) or plasmapheresis.

Why would I have therapeutic plasma exchange?

Therapeutic plasma exchange may be recommended to:

- Stabilize a rapid decrease in muscle strength, including significant difficulties in speaking, swallowing and breathing
- Optimize MG status before surgery
- Add to present treatment if current forms of therapy are providing insufficient disease control

How does therapeutic plasma exchange work in MG?

The process removes the antibodies that are interfering with function of the neuromuscular junction. Removal of these pathogenic antibodies allows for restoration of normal activity at the neuromuscular junction, resulting in improved muscular strength.



How many therapeutic plasma exchanges will I need and where will it be done?

The number of treatments depends on whether it is for preventative treatment or for a crisis.

Preventative Treatment: Your neurologist will work with you and the apheresis team to determine the number of treatments needed based on your MG status and signs/symptoms. Treatment options vary from every 24 to 72 hours for three to six treatments, or single treatments weekly, bi-weekly or monthly until signs and symptoms diminish.

Crisis: If experiencing an MG crisis, therapeutic plasma exchange can significantly improve signs and symptoms. The health care team will assess your neurologic status, muscular strength, and vital signs to assist in determining the number of treatments needed. In general, a patient with MG crisis receives a more aggressive therapeutic intervention and will have therapeutic plasma exchange procedures every 24 to 48 hours for 5 to 6 treatments until you are stable.

Therapeutic plasma exchange can be administered both in the hospital and in an outpatient setting. The procedure can be completed by two different delivery systems:

- Peripheral venous access if you have suitable arm veins.
- Placement of a double lumen central venous catheter or subcutaneous port. Overnight hospitalization is sometimes recommended for central venous catheter placement and initiation of therapeutic plasma exchange. Generally, a tunneled catheter or port is needed to minimize risk of line infections if long-term therapeutic plasma exchange treatment is anticipated and peripheral venous access is not possible.

What type of venous access is required?

In the case of therapeutic plasma exchange using peripheral veins, a needle will be inserted into a vein in each arm and removed after each treatment. In rare instances, only one arm may be used. If your arm veins are unsuitable to use, interventional radiologists or surgeons will place an indwelling, double lumen catheter into a large vein in your neck or shoulder under local anesthesia. This is also known as a central catheter or central line. Such indwelling catheters can remain in place for many months with proper care. In rare, urgent situations, catheters are placed in the groin, but these are temporary due to high risk of infection and because they interfere with

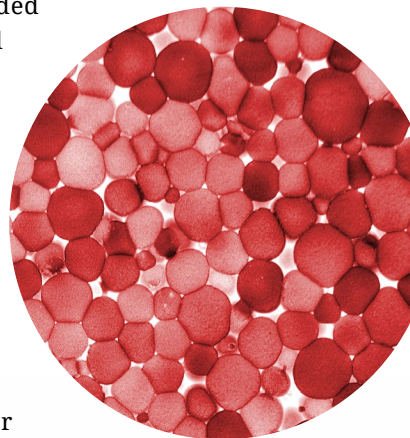
walking. They are usually reserved for inpatient therapeutic plasma exchange. You will receive instructions from your team to prepare for indwelling catheter placement. Be aware of the difference between internal and external catheters when showering and



submerging under water. Maintenance and proper cleaning is extremely important to avoid risk of infection.

Another option for long-term vascular access is a subcutaneous port. These devices are completely embedded under the skin and so have lower risks of infection and incidental damage compared to catheters.

However, a port requires a slightly larger procedure during placement and is not ready for immediate use; thus, this device may or may not be a suitable vascular access option depending on your specific clinical situation.



What should I expect during therapeutic plasma exchange?

Unless the physician has instructed otherwise, it is important to eat before the therapeutic plasma exchange and not skip meals. During therapeutic plasma exchange, you may drink fluids. You should empty your bladder prior to the procedure and ask the nurse for a bedpan or urinal if needed during the therapeutic plasma exchange procedure. If you have peripheral venous access, you will need to keep both arms outstretched and still. If you have an indwelling catheter, you will be able to move your arms during the procedure.

Wearing comfortable clothing with loose fitting sleeves that pull easily above your elbows will make it easier to place the needles in each arm for peripheral access. Wearing comfortable



clothing with a loose-fitting shirt is important if you have an indwelling catheter to provide easy access to the site. Bringing something to read, watch, or listen to will help you pass the time. You may experience

some coldness during the treatment. If this happens, ask your treatment staff to provide you with a warm blanket.

How long does therapeutic plasma exchange take?

The time spent on the machine may take from one to three hours. This depends upon many factors such as your weight, height, the amount of plasma to be exchanged, the catheter type (peripheral or central), and catheter performance.

How will I feel after my therapeutic plasma exchange?

Many patients feel fine after the procedure. Others may feel tired or have lightheadedness. If the procedure is done on an outpatient basis, then someone should drive you home.

How soon will the treatment work?

Compared to other treatments, therapeutic plasma exchange works quickly to increase strength. You are likely to notice improvement after the first few procedures.

How long will the improvement last?

Effectiveness can vary, but most people who have received five therapeutic plasma exchanges over 1 to 2 weeks can remain stronger for one to

two months. After that time, weakness can recur due to the pathogenic antibodies returning to pretreatment levels in the bloodstream and at the neuromuscular junction.

What are the possible adverse effects?

Common adverse effects may include a change in heart rate or blood pressure, sweating, or feelings of faintness, dizziness, coldness, or abdominal cramps. Additionally, tingling associated with the mouth, eyes, fingers or toes can occur. Rare, adverse effects may include bleeding (a result of removal of clotting factors) and a possible allergic reaction such as itching, wheezing, and rash to the solutions used during the procedure. Infection or clotting can occur with vascular access devices placed for therapeutic plasma exchange, which often then necessitates removal of the device. Most individuals tolerate therapeutic plasma exchange very well and have no issues during their treatments.

I am anxious about having therapeutic plasma exchange!

It is normal to feel nervous about any new procedure. Anxiety about therapeutic plasma exchange – especially during the first few treatments – is only natural and can produce hypersensitivity to new sensations that will go away once you become more comfortable with the procedure. Helpful ways to prepare and alleviate fears may include speaking to someone who has been treated with therapeutic plasma exchange and discussing



concerns with your physician and the apheresis team. It can help to ask the team to explain what they are doing and why during the procedure. Contact MGFA if you would like to connect with someone else with MG who has experience with therapeutic plasma exchange.

I am a family member/friend of a person with MG, what do I need to know?

It can be very comforting for a person going through therapeutic plasma exchange to have someone there as company, and to ask about what is happening through the procedure. In preparing for possible emergencies, it is important to know that therapeutic plasma exchange (or IVIg) may be started right away for a person with MG who is in full-

blown crisis — having difficulty swallowing, speaking,

breathing, and even

needing assistance with respiration using a mechanical ventilator or other device. This can be scary for everyone, especially for YOU as you find yourself having to take charge of working with the health care team.

The number of therapeutic plasma exchange treatments will vary, however, it is crucial

to know what to expect. What you are seeing on the monitor (heart rates, respiration rates, blood pressures, et cetera), and the role mechanical ventilators and other devices play in the patient's recovery. It can be intimidating to see your family member connected to a machine, but it helps to know this process may

be the fastest way for a person with MG to regain strength. Be prepared for breathing trials that can be uncomfortable for your loved one, but are important tests to determine their strength prior to removing any breathing device.

Speak to your loved one regularly; explain what is happening and sound as calm and confident as you can. Even if he/she is unable to answer or is sedated, hearing your voice can be very soothing. In your role as advocate, ask questions about what you do not understand so you can be at ease about your loved one's care.

Remember to take care of yourself throughout a crisis as you will play a key role in the days and weeks to come. The real work begins when the patient is breathing without any assistance and coping with everything that just transpired. You will need to be both physically and mentally strong for your loved one.





Myasthenia Gravis Foundation of America

Our Vision: A World Without MG

Our Mission: Create Connections, Enhance Lives,
Improve Care, Cure MG

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