

THYMECTOMY

Thymectomy

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**Common questions patients
ask about thymectomies.**

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The following are some of the most common questions asked when a thymectomy is being considered for adult and younger patients with autoimmune myasthenia gravis (MG).

The answers supplied below are presented in general terms as background information only and should not be used to make specific decisions. Since each patient's situation is unique and the types of thymectomy being performed vary, it is essential to discuss in detail these and all other questions about the surgery with the MG specialist and the surgeon.

What is a thymectomy and why is it performed?

A thymectomy is the surgical removal of the thymus gland. The thymus has been demonstrated to play a role in the development of MG. It is removed in an effort to improve the weakness caused by MG, and to remove a thymoma if present.

About 10% of MG patients have a tumor of the thymus called a thymoma. Most of these tumors are benign and tend to grow very slowly; on occasion they are malignant ("cancerous").

What is the function of the thymus? Is its removal harmful?

The thymus plays a major role in the development of the body's immune system. This function appears virtually complete by birth. Removal of the thymus in the treatment of MG does not affect the immune system thereafter.

Exactly where is the thymus located?

The thymus is located in the front portion of the chest (anterior mediastinum) with "finger-like" extensions into the neck and consists of multiple lobes (two to five or more). In addition, varying amounts of thymic tissue may be present in the fat surrounding the lobes, both in the neck and chest.

Who should have a thymectomy?

Although it is not definitely established which patients should have a thymectomy or what type of operation should be performed, a thymectomy is frequently recommended for patients under the age of 60 (occasionally older) with moderate to severe MG weakness. It is sometimes recommended for patients with relatively mild weakness, especially if there is weakness of the respiratory (breathing) or oropharyngeal (swallowing) muscles. It is also recommended for all patients with a thymoma. A thymectomy is usually not recommended for patients with weakness limited to the eye muscles (ocular myasthenia gravis).

What should I expect as I consider a thymectomy?

When a thymectomy is being considered, the patient is referred to a surgeon. It is important to choose a surgeon experienced in performing thymectomies for patients with MG. The surgeon will review the clinical records, examine the patient, discuss the surgical choices with the patient and make a recommendation. The surgeon also explains the anticipated pre- and post-operative courses, possible complications, and the anticipated results. The patient, in consultation with the neurologist and surgeon, then makes a decision whether to proceed with a thymectomy and the type of surgery to be used.

What are the goals of a thymectomy?

The neurological goals of a thymectomy are significant improvement in the patient's weakness, reduction in the medications being employed, and ideally eventually a permanent remission (complete elimination of all weakness off all medications).

A thymectomy is not used to treat active disease but rather it is believed to improve long-term outcome. This may not be seen for one-two years or more. A research trial is currently underway (partially funded by MGFA) to examine the effects of thymectomy on MG patients. These operations are now performed with minimal risk.

How is the surgery performed?

There are three basic surgical approaches, each with several variations. Regardless of the technique employed, the surgical goal is to remove the entire thymus. Many believe this should include removal of the adjacent fat; others are less sure.

Transsternal Thymectomy

Incision: Vertical (lengthwise) on the anterior chest; the sternum (breastbone) is “split” vertically.

Thymus Removal: The chest and neck portion of the thymus are removed through this incision.

Extended Form: The fat located in the front part of the chest next to the thymus, as well as the thymus, is removed. Complete removal of all tissue containing thymus is believed ensured.

Combined Chest and Neck: A few MG Centers add a formal neck dissection to the sternal technique to also ensure the removal of all the thymus in the neck.

Thymoma: Most recommend the transsternal approach for removal of a thymoma.

Transcervical Thymectomy

Incision: Transverse (horizontal) across the lower neck.

Thymus Removal: The chest portion of the thymus is removed through this incision.

Extended Form: The “extended” form allows improved exposure of the thymus in the chest with more complete removal of the thymus. Although the adjacent fat is also removed, less is removed than in the extended transsternal thymectomy.

Videoscopic (VATS) Thymectomy

Incision: Several small incisions on the right or left side of the chest.

Thymus Removal: Fiber-optic instruments are used. These are small flexible tubes with a light at the end through which small instruments can be passed. The amount of thymus and fat removed is variable.

Extended Form: In the “VATET” form, incisions are made on both sides of the chest, as well as in the neck, for “more complete” removal of the thymus.

What are the results of a thymectomy?

Many neurologists experienced in the treatment of MG are convinced that a thymectomy plays an important role in the therapy of MG, although the benefit is variable, hard to define and remains unproven by today’s strict standards.

In general, most patients begin to improve within one year following a thymectomy and a variable number eventually develop a permanent remission (no weakness and no medication). Some physicians believe the remission rates

after surgery are in the 20-40% range regardless of the type of thymectomy performed. Others believe that the remission rates following the more extensive procedures are in the 40-60% range five or more years after the surgery.

It is important to note that rigorous scientific studies are needed to resolve the debate concerning the role of and best method of performing a thymectomy in the treatment of MG patients.

What type of thymectomy should I have?

Since there is no universal agreement, or unequivocal proof, as to which type of thymectomy is best, it is difficult for patients to decide what is best for them. There is, however, general agreement that the entire thymus should be removed and that the patient should select the procedure that ensures as much as possible that this is accomplished. Some surgeons believe that all the surrounding fat should be removed as well, because it frequently contains microscopic (very small) amounts of thymus; others believe this may not be necessary.

The most frequently used procedure is the extended form of the transsternal thymectomy. Its proponents believe that it gives the best assurance that, in most instances, the entire thymus is being removed and performed safely, and that it produces the best long-term results.

Those advocating the transcervical or the videoscopic thymectomy do not share the above observations. They believe the “minimally invasive” procedures (transcervical and videoscopic) are as effective.

Since there is no absolute proof as to which type of thymectomy is the procedure of choice, patients need to be fully informed, review the evidence presented by the neurologist and surgeon caring for them, and perhaps obtain additional consultation.

What can the MG patient expect in the pre-operative, anesthesia and post-operative periods?

In general, MG Centers have developed protocols for the care of MG patients and have a team of neurologists, surgeons, pulmonologists, intensive care and respiratory care specialists, nurses and anesthesiologists caring for MG patients undergoing a thymectomy. Patients should discuss all aspects of the pre-and post-operative care and anesthesia with the surgeon, anesthesiologist and neurologist.

To reduce the risks of post-operative respiratory complications or the post-operative need for prolonged respiratory support with a ventilator (breathing machine), many patients require pre-operative plasma exchange or intravenous immunoglobulin (IVIg), and some require immunosuppressive therapy as well. Pyridostigmine, if being administered, may or may not be discontinued the day of surgery, and may or may not be restarted immediately post-operatively.

The anesthesia for patients with MG is similar to the anesthesia given to other patients. An endotracheal tube (tube in the windpipe) is inserted after the patient is asleep. Muscle relaxing drugs, however, are usually avoided. The patient may or may not be extubated (removal of the endotracheal tube) upon awakening, depending on the patient's strength. If the endotracheal tube is not removed on awakening, the tube will be attached to a ventilator.

Ordinarily after the surgery, the patient will go to a Recovery Room, Respiratory Care Unit or Intensive Care Unit depending on each hospital's method of taking care of MG patients following surgery. A ventilator may be required depending on the type of operation and the severity of the patient's weakness. As soon as the breathing tube has been removed, the patient will be asked and helped to deep breathe and cough frequently to keep the lungs clear of secretions. One or two chest tubes (small tubes exiting the chest and attached to drainage bottles) are usually used after the transsternal and videoscopic operations, and removed soon after surgery.

Medications used to manage MG before surgery are usually continued after surgery. The neurologist will decide how to taper your medications after surgery in subsequent follow-up appointments.

Pain is minimal following transcervical thymectomy and usually mild following videoscopic thymectomy, although some patients have reported late pain. The pain associated with transsternal thymectomy is temporary, well controlled with medication and gradually resolved within 3-5 days. Patients typically require minimal pain medication on hospital discharge.

The length of time in the hospital will vary depending on the type of surgery and the patient's overall weakness. In most cases the patient will be ready to go home in a few days to a week. The patient's preoperative medications,

immunosuppression and other forms of therapy are usually resumed after surgery for variable periods of time depending on the MG symptoms and the neurologist's recommendations.

When can I return to my usual activities?

Patients should discuss this with the surgeon in advance and let their employer or school know the anticipated time they will miss.

The recovery period and the time away from regular activities, like work or school, will vary depending on the patient's weakness, type of surgery and type of the patient's activities. A patient who does heavy lifting or construction work will be off work longer than someone who has a desk type job. In general, three to six weeks of limited activities is a common length of time for recovery.

Will my insurance pay for the surgery?

Health insurance companies do pay for thymectomies. Since there may be questions concerning insurance coverage with specific surgeons and specific institutions, it is a good idea to check with your insurance company as soon as a thymectomy is being considered to make sure that the coverage is in order. Some insurance companies may require additional consultation and some type of prior authorization. In some instances letters from the neurologist and surgeon will be required to defend the procedure selected.

These are just a few of the many questions patients with MG may have while considering a thymectomy. Physicians and nurses want their patients to do well, and part of that consists of patients being as informed and involved with their own care as possible.

It is encouraging to note that most patients have very few problems after the surgery and are able to return to their usual pre-operative activities relatively soon.

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