*Original Article*

# Surgery of Myasthenia Gravis Associated or Not With Thymoma: A Retrospective Study of 43 Cases

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***Objectives:*** **Thymectomy is a surgical treatment of myasthenia gravis. Our goal is to report our experience in the surgical treatment of myasthenia gravis with or without thymoma and a review of the literature.**

***Materials and Methods:*** **This is a retrospective study over a period of 10 years (2001–2010) on 43 patients: 28 women and 15 men with a mean age of 39.3 years (range 16–68 years). The myasthenia gravis was confirmed by clinical, electromyographic data and the presence of antibodies to acetylcholine receptors.**

***Results:*** **Computed tomography objectified thymic mass in 14 cases (32.5%) enlarged thymus without visible mass in eight cases (18.6%). All patients received anticholinesterase, cortico steroids in 25 cases and in three cases plasmapheresis was required. The surgical approach was total sternotomy ($n = 32$ cases), cervicotomy ($n = 2$), cervical and manubriotomy ($n = 1$), a manubriotomy ($n = 3$) and a thoracotomy in five cases (lateralised thymoma). All patients underwent a total thymectomy associated or not with resection of the tumour. Intensive Care Unit was necessary for at least 24 h up to six days. The postoperative course was marked by a myasthaenic crisis ($n = 2$) and respiratory failure ($n = 3$) with a favourable outcome. The prognosis was marked by a complete remission in 14 cases, partial remission in 11 patients, stabilisation ($n = 16$ cases) and increasing crisis in two patients.**

***Conclusion:*** **Thymectomy certainly allows clinical improvement and reduced crisis of myasthenia gravis. Long term monitoring will confirm the benefit of non-oncological thymectomy alone or in combination with standard treatments for patients with generalised myasthenia gravis without thymoma.**

**(Heart, Lung and Circulation 2013;22:738–741)**



**Keywords. Myasthenia gravis; Thymoma; Thymic hyperplasia; Sternotomy**

## Introduction

Myasthenia gravis (MG) is an uncommon autoimmune disease, characterised by circulating antibodies directed against the nicotinic acetylcholine receptor in the neuro muscular junction, which explain the pathogenesis of muscle fluctuating weakness, worsening with exertion, and improving with rest. The severity of the disease depends on the involvement of respiratory muscles, other bulbar muscles and limb musculature. The new treatment modalities allowed a significant reduction in morbidity and mortality. Surgery has a place in the treatment of MG with or without thymoma.

## Materials and Methods

From January 2001 to December 2010 a retrospective study included 43 patients who underwent thymectomy for MG with or without thymoma. Our descriptive analysis allowed an evaluation in the short and medium term of the role of surgery as a therapy in MG.

## Results

The average age was 39.3 years (range 16–68 ans) with a female predominance: 28 women (65.1%) and 15 men (34.9%). We noted chronic smoking in four cases, diabetes ($n = 7$), hypertension ($n = 2$) and pulmonary tuberculosis in one case.

Patients were classified clinically according to the classification MGFA (Myasthenia Gravis Foudation of America):

stage **I**: 21 patients, **IIA**: 12 patients, **IIB**: 04 patients, **III**: 03 patients and **IV**: 03 patients.

Received 9 December 2012; received in revised form 8 February 2013; accepted 28 February 2013; available online 30 March 2013

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1443-9506/04/$36.00
http://dx.doi.org/10.1016/j.hlc.2013.02.010

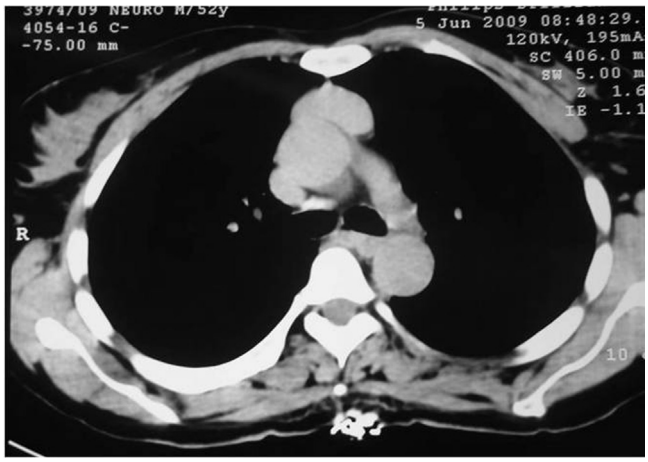


**Figure 1.** *CT scan showing a well circumscribed anterior mediastinal mass tissue slightly lateralised to the right side.*

Explorations include the electromyogram that objectified reduced neuromuscular transmission and postsynaptic block in all cases.

- The immunological test and measures of anti-AChR antibodies was positive in 42 patients except for one case where it was normal with positive Anti MuSK.
- Cervical and chest computed tomography, objectified thymic mass in 14 cases (32.5%) (Fig. 1), an enlarged thymus without individualised mass in eight cases (18.6%) and was normal in 21cases (48.8%). Thymic calcifications were observed in two patients (Fig. 2).

All patients received medical treatment by anticholinesterase. Clinical improvement was totally noted in 16 patients, partial in eight patients. Corticosteroid therapy was necessary in 25 patients and three patients needed plasmatic exchange and intravenous immunoglobulin.

Surgical approach was total median sternotomy ($n = 32$ cases), cervicotomy ($n = 2$), cervicomanubriotomy ($n = 1$), a manubriotomy ($n = 3$), and posterolateral thoracotomy in five cases (lateralised thymoma).

All patients underwent a total thymectomy (Fig. 3A) associated with removal of probably existant thymoma (Fig. 3B). We always ensure thymic and pericardial fat resection (Fig. 3C).

The pathologist confirmed thymoma in 14 cases (32.5%), thymic hyperplasia in eight cases (18.6%) and normal thymus in 21 cases.

Postoperative course was uneventful in 38 patients. In five cases complications occurred: typical myasthenic crisis ($n = 2$) and respiratory failure ($n = 3$). ICU was necessary three to eight days. One patient presented pericardial effusion 35 days after surgery and percutaneous pericardiocentesis allowed good evolution.

The outcome after 12–36 months (with an average of 21.6 months) was characterised by: complete remission ($n = 14$ cases), partial remission in 11 patients, stabilisation of MG ($n = 16$) and increasing symptoms in two patients.

## Discussion

Myasthenia gravis is the most common disease of the neuromuscular junction, but remains relatively rare. Its incidence varies from 1.7 to 10.4 per million per year. However, the authors agree on the existence of two frequency peaks: the first peak between 20 and 40 years where women are preferentially affected and a second after 50 years where men are more often affected [1]. The role of the thymus in the pathogenesis has been well established by producing circulating antibodies against postsynaptic acetylcholine receptors [2].

The treatment of MG is based on optimising neuromuscular transmission and reducing pathogenic antibodies. It includes symptomatic and immunomodulatory therapy [3].

In the absence of established guidelines, proposals from the literature and practical experience may be issued.

Strategy management of patients with MG is always guided by several parameters: age, seat of deficits, social and professional impact of the disease, associated diseases, presence or absence of a thymoma. Medical treatment is based first on anticholinesterases, corticosteroids started gradually according to clinical response [4], and immunosuppressors [5].

The efficacy of thymectomy is still controversial. Surgical treatment is strongly recommended for patients with thymoma. Thymectomy is advised as soon as the patient's degree of weakness is sufficiently controlled to permit surgery.

Most authors accepted two strategies [6]: MG with thymoma and MG without thymoma. We agree with this subdivision.

(1) *Thymoma with myasthenia*: surgery is oncological. Thymectomy must be complete as much as possible. The approach is sternotomy. Other incisions are available such cervicotomy, mediastinoscopy or thoracoscopy. Thoracotomy if the tumour is lateralised, but must allow complete thymic resection. The prognosis depends on the severity of the MG, the stage of tumour extension, completeness of resection, pathological type. This later is a relatively new concept and requires probably to be confirmed [7]. In a recent study of 228 Chinese patients the prognosis of thymoma associated with myasthenia is similar to that which is not associated with MG [8]. The risk of local recurrence requires regular monitoring by chest CT.

(2) *Myasthenia without thymoma*: despite indications and statements from leading societies, such MGFA and International Thymic Malignancy Interest Group (ITMIG), there are a lot of controversial issues and grey areas which need more collaborative studies. Nowadays a controlled clinical trial of five years (MGTX study) with two groups (one group receiving corticosteroids alone and a group treated with corticosteroids and thymectomy) is carried out to investigate the impact of thymectomy in non oncological myasthenia [9].