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Outcomes of thoracoscopic thymectomy in patients with juvenile myasthenia gravis☆

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ABSTRACT

Introduction: Myasthenia gravis (MG) is an autoimmune disorder of the postsynaptic neuromuscular junction resulting in fatigability of voluntary muscles. There has been increasing evidence supporting thymectomy for MG in adults, and evidence for the role of surgery in pediatric age groups is increasing. The purpose of this study is to describe the outcomes of our patients with juvenile MG undergoing thoracoscopic thymectomy.
Material and methods: All patients with juvenile MG who underwent thoracoscopic thymectomy at Phoenix Children's Hospital between 1999 and 2014 were included. Patients were diagnosed by their treating neurologist. An Osserman and Genkins criterion was used to classify the severity of the disease and DeFilippi classification was used to assess remission.
Results: Twelve patients underwent thoracoscopic thymectomy for juvenile MG during the time frame studied. Nine (75%) patients had an Osserman stage of IIB, with only two patients with ocular disease. There were no conversions to an open procedure. Seven (59%) patients had normal thymic histology, 4 (33%) had evidence of follicular hyperplasia and one (8%) had involutional changes. The median length of hospital stay was 2 days (range 1–5 days). There was no 30-day postoperative morbidity, reoperations or mortality. The median length of follow-up was 31 months (range, 4–91 months) and at the time of their last follow-up; all 12 (100%) patients had a DeFilippi Classification of 3 or better.
Conclusion: Surgery for MG in children is indicated for antibody-receptor-positive patients with moderate to severe disease. Thoracoscopic thymectomy is a safe and acceptable treatment for juvenile MG with good disease control. The low morbidity and shorter hospital duration make it an excellent option for consideration.

Myasthenia gravis (MG) is an autoimmune disorder of the postsynaptic neuromuscular junction resulting in fatigability of voluntary muscles. A recent review showed an incidence of MG for the population in Europe to be approximately 30 per million per year, with an incidence in children and adolescents aged 0 to 19 being between 1 and 5 per million per year [1]. Thymectomy for MG is becoming standard treatment in non-muscle-specific tyrosine kinase, acetylcholine-receptor-antibody-positive MG with generalized symptoms. Timing of surgical intervention for MG lacks standardization and has often been a subjective decision based on the risk-to-benefit ratio of undergoing thymectomy [2].

The thoracoscopic approach was initially described in the adult literature [3,4]. Increasing evidence exists supporting thymectomy for MG in adults [5–7], and the role of surgery in pediatric age groups has been supported by several small series [1,2,8–10]. Additionally, a few small series have shown that a thoracoscopic approach is safe, with reasonable improvements in postoperative outcomes [8,11]. To date, only two studies [2,9] have compared different surgical approaches in patients with juvenile MG and both were limited owing to small sample size and restricted follow-up.

Due to the relative paucity of juvenile MG, there are no large studies of thoracoscopic thymectomy in children. The purpose of this study is to describe the outcomes of our relatively large series of patients with juvenile MG undergoing thoracoscopic thymectomy and to support the growing body of evidence in favor of it.

1. Materials and methods

All patients with juvenile MG who underwent thoracoscopic thymectomy at Phoenix Children's Hospital between 1999 and 2014 were included. Study approval was obtained from the Institutional Review Board; informed consent was waived. The medical records were reviewed for patient demographics, preoperative symptoms and medical treatment, preoperative imaging including computed tomography or magnetic resonance imaging of the chest, operative data including details of surgery, estimated blood loss, intraoperative complications, and postoperative data including length of intensive care unit stay, hospitalization, intubation and any complications.

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Group I	**Ocular symptoms**
Group IIA	**Mild, generalized symptoms (including bulbar)**
Group IIB	**Moderate, generalized symptoms**
Group III	**Acute, fulminating symptoms**

Fig. 1. Disease classification, Osserman and Genkins criteria [11].

Patients were diagnosed with MG by their treating neurologist based on history, physical examination, positive response to anticholinesterase agents, and positive acetylcholine receptor antibodies. Osserman and Genkins criteria were used to classify the severity of the disease preoperatively [12] (Fig. 1). Status of the disease postoperatively was assessed by the DeFilippi classification at their last follow-up [13] (Fig. 2). Both preoperative and postoperative classifications were determined by the same treating neurologist.

1.1. Surgical technique

A right-sided thoracoscopic approach is performed in all of our patients undergoing thymectomy. Early in the series, the patients were positioned supine although currently the torso is also elevated using gel rolls. The table is rotated right-side up at the start of the procedure. This position gives unrestricted access to chest from posterior axillary line forward, avoids instrument restriction by the excessive table width (in small children), and provides the option for bilateral thoracoscopy, if needed (although not required yet). The right side also provides excellent access to the thymus near the superior vena cava, as well as where the right phrenic nerve courses caudally. An initial 5 mm anterior axillary line incision is made and pleural cavity entered using optical guidance. An insufflation pressure of 8 mmHg was maintained for the entire procedure. Two subsequent 5 mm ports are placed, at the midclavicular inframammary line and posterior axillary line, at the 4th intercostal space (Fig. 3).

The pleura is incised along the right side of the thymus and dissected off the pericardial sac and aorta. Hook cautery is used for most of the dissection. Dissection is then carried cephalad into the neck while taking care to avoid injury to the right phrenic nerve which courses very close to the thymus near the superior vena cava. The venous drainage to the brachiocephalic vein is ligated using LigaSure (Medtronic, Minneapolis, MN). Dissection is then continued from right to left to peel the thymus from the left pleura. Next, we direct our attention cephalad similar to the right side and dissect the upper pole off its attachments, again ligating major venous branches with a LigaSure. The posterior port is enlarged to a 12-mm port; the thymus is retrieved using an endoscopic retrieval bag.

1.2. Statistical analysis

Descriptive statistics were computed using SPSS version 16.0 (Armonk, NY). Continuous variables (age, duration of surgery, length of follow-up) are reported as median (range). Categorical variables (e.g. preoperative testing, disease classification) are reported as frequencies and percentages.

2. Results

2.1. Demographics and preoperative characteristics

A total of twelve patients underwent thoracoscopic thymectomy for juvenile MG during the time frame studied. Seven (58%) patients were males with the median age of 11 years (range, 3–17). Mean duration between diagnosis and surgery was 418 (75–1756) days. All patients were diagnosed by either a positive edrophonium test or a positive acetylcholine receptor antibody. Nine (75%) patients had an Osserman stage of IIB, with only two patients with ocular disease (Stage I) (Table 1). All patients underwent preoperative medical treatment and required intravenous immunoglobulin. One patient had an admission fora myasthenia crisis and required plasmapheresis before undergoing surgery.

2.2. Perioperative characteristics

All operations were done thoracoscopically without any conversion to open procedure. There was minimal blood loss during the procedure with no patient requiring a blood transfusion, either intra- or postoperatively. A total of 9 patients (75%) had a chest tube inserted at the time of surgery; all of them on the right side and all removed by postoperative day two (Table 2).The chest tube was more commonly left early in the

Class I	**Complete remission, no medication requirements**
Class 2	**Asymptomatic; decreased medication requirements**
Class 3	**Improvement in symptoms; decreased medication requirements**
Class 4	**No change in symptoms or medication requirements**
Class 5	**Worsening symptoms**

Fig. 2. DeFilippi classification of remission [12].