Video-assisted thoracoscopic extended thymectomy in myasthenic children

Rita Sonzogni,1 Lorenzo Novellino,2 Alberto Benigni,1 Ilaria Busi,1 Magda Khotcholava,1 Angelica Spotti,1 Valter Sonzogni1
1Department of Anesthesia and Intensive Care I, A.O. Ospedali Riuniti di Bergamo; 2Department of Surgery, A.O. Bolognini di Seriate, Italy

Abstract

Myasthenia gravis (MG) is an autoimmune disease marked by weakness of voluntary musculature. Medical and surgical therapy of adult myasthenia is well documented. There is little pediatric surgical evidence, only a few case reports being available. The aim of this paper is to verify whether the surgical and anesthesiological techniques can warrant an early and safe discharge from the operating room. The secondary aim is to assess the presence of perioperative indicators that can eventually be used as predictors of postoperative care. During the years 2006-2009, 10 pediatric patients were treated according to a surgical approach based on video assisted thoracoscopic extended thymectomy (VATET). Standard preoperative evaluation is integrated with functional respiratory tests. Anesthetic induction was made with propofol and fentanyl/remifentanyl and maintenance was obtained with sevoflurane/desflurane/propofol ± remifentanyl. A muscle relaxant was used in only one patient. Right or left double-lumen bronchial tube (Rüsch Bronchopart® Carlens) placement was performed. Six patients were transferred directly to the surgical ward while 4 were discharged to the intensive care unit (ICU); ICU stay was no longer than 24 h. Length of hospital stay was 4.4±0.51 days. No 4 were discharged to the intensive care unit (ICU); ICU stay was no longer than 24 h. Length of hospital stay was 4.4±0.51 days. No patient was readmitted to the hospital and no surgical complications were reported. Volatile and intravenous anesthetics do not affect ventilator weaning, extubation or the postoperative course. Paralyzing agents are not totally contraindicated, especially if short-lasting agents are used with neuromuscular monitoring devices and new reversal drugs. Perioperative evaluation of the myasthenic patient is mandatory to assess the need for postoperative respiratory support and also predict timely extubation with early transfer to the surgical department. Availability of new drugs and of reversal drugs, the current practice of mini-invasive surgical techniques, and the availability of post anesthesia care units are the keys to the safety and successful prognosis of patients affected by MG who undergo thymectomy.

Introduction

Myasthenia gravis (MG) is an autoimmune disease marked by weakness of voluntary musculature. Although it can involve all the striate musculature of the organism, it mainly affects muscular groups innervated by brainstem motor nuclei (eye and eyelid muscles, masticatory and swallowing muscles, mimic musculature). Neuromuscular transmission can be compromised in three different ways: i) the antigen-antibody complex formation can be blocked; ii) complement-mediated destruction of postsynaptic receptors; iii) elevated receptor degradation ratio. From a biological point of view, specific antibodies against nicotinic receptor of acetylcholine (AChR-Ab) can be found in 85-95% of cases with a generalized presentation of the disease, while only 60% of ocular presentation of the disease shows this antibody.

MG is included in the group of neuromuscular diseases and junctional myopathies. There are some specific subgroups: newborn presentation, congenital presentation or the more frequent early presentation. Diagnosis can be complex and is not only clinical but based on the serum finding of AChR-Ab and muscle-specific kinase receptor antibodies (anti-MuSK). Edrophonium tests can also be used, such as electromyography and chest X-rays.

The incidence of MG is 150-400 new cases per million per year. There is a female prevalence in early presentation and a male prevalence in presentation in the patient’s 50s and 60s.

Medical and surgical therapy of adult myasthenia is well documented while pediatric surgical evidence is very poor and only a few case reports are available.1,3 The causes of this can be identified in the less frequent pediatric presentation, lack of availability of centers dedicated to pediatric MG, while both adult and pediatric patients are collected in the same centers.

Surgery offers a real chance of improvement in MG and although not a radical therapy, perioperative evaluation of the myasthenic patient (neurological functional capacity, grade of bulbar involvement, respiratory function) is mandatory to assess the need for postoperative respiratory support and also predict timely extubation with early transfer to the surgical department.4,5

In this setting, myorelaxants are used less during anesthesia because of their unpredictable response, and preoperative sedation is also avoided in order to lower the risk of aspiration, especially in patients with reduced respiratory reserve.5,7 Therefore, the main aim of this paper is to verify whether the surgical and anesthesiological techniques can ensure an early and safe discharge from the operating room leading to immediate discharge to the surgical ward.

The secondary aim is to assess the presence of perioperative indicators that can eventually be used as predictors of postoperative care.

Materials and Methods

This study was carried out from 2006 to 2009 at a reference institution for surgical treatment of MG. The surgical approach is based on video assisted thoracoscopic extended thymectomy (VATET). VATET was developed in the 1990s and is a mini-invasive option for thymectomy. The incision is 3-4 cm in length and is made through the midclavicular line, below the clavicle, and above the sternum. The incision is carried down to the level of the pericardium, and the pericardium is separated from the sternum. The thymus is then dissected from the pericardium, and the thymectomy is performed. The thymus is then removed from the chest and sent for pathological examination. The sternum is then closed, and the incision is closed in layers. The patient is then extubated and transferred to the intensive care unit. The patient is then monitored for 24-48 hours, and then discharged home.

Correspondence: Rita Sonzogni, Department of Anesthesia and Intensive Care I, A.O. Ospedali Riuniti di Bergamo; via Mazzini 4, 24030 Mozzo (BG), Italy. Tel. +39.3470479323. E-mail: sonzogni.r@gmail.com

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and ascending aorta from heart to right ano-
ymous vein). There is then a third left thoraco-
oscopic surgery that permits full dissection.
Finally, the extraction of the thyme from the
thorax is made from a cervicothomic access.
The need for surgical therapy is obviously
assessed by the neurologist. Medication is
stopped the day of surgery and restarted some
hours after discharge to the ward, following
further neurologist referral.
Standard preoperative evaluation with full
blood tests, echocardiogram (ECG), chest X-
ray (CXR) is integrated with functional respi-
atory tests (spirometry, blood gas analysis).
The severity of the disease and evaluation of
clinical stability are assessed by the neurolo-
gist using the MG Foundation of America
(MGFA) scale, as described in Table 1.

Results

During the years 2006-2009, 10 pediatric
patients from 9 to 17 years old (9 females, one
male) were treated. Patients’ characteristics
are described in Table 2.
Diagnosis was at least two years before
treatment and all patients were treated with
steroids and cholinesterase inhibitors. Anti-
AChR antibody titters were positive in 8 cases,
and anti-MUSK in one. Only one patient was
seronegative. No preoperative myasthenia cri-
sis was detected.
Nine of the patients were defined to be in
good clinical condition and only one was in
partially satisfactory condition (MGFA Class
III).
Blood gas analysis was normal in all
patients while functional respiratory tests
were normal in 6. One patient showed restric-
tive respiratory syndrome, while one patient
showed obstructive respiratory syndrome. Gas
diffusion capability was mildly altered in one
patient.
Two patients received preoperative sedation
with benzodiazepine. Anesthetic induction
was made with propofol 2.5-3.5 mg/kg and fen-
tanyl 2 mcg/kg (6 cases) or propofol 2.5-3.5
mg/kg and remifentanil 0.05-0.2 mcg/kg/min
(4 cases). Anesthetic maintenance was
obtained with sevoflurane (4 cases), sevoflu-
trane/remifentanil (4 cases), propofol/remifen-
tanyl (one case) and desflurane/remifentanil
(one case). A muscle relaxant (cisatracurium
0.1 mg/kg) was used as induction in only one
patient (who was later one of those to be trans-
ferred directly to the surgical ward). In this
case, Train of Four monitoring (TOF) was
used.
The radial artery was cannulated in order to
measure invasive blood pressure; intra-opera-
tive monitoring consisted in internal tempera-
ture, functional respiratory data, end-tidal car-on dioxide (ETCO2), oxygen saturation
(SpO2) and ECG. Left DLT (Rüsch
Bronchoparth® Carlens) was placed in 9
patients, a right DLT was placed in one case;
sizes ranged from 32 to 35.
The average surgical time was 123±18.9
min, the average weight of thymus with fat tis-
sue was 49.8±4.4 grams, the presence of thy-
moma was established in 2 cases and of thymic
hyperplasia in the other 8.
Operating room (OR) discharge was
40±18.5 min after awakening (cumulative
time for both transfer to the surgical ward or
ICU). Six patients were transferred directly to
the surgical ward. Four were discharged to the
ICU, one of whom on the specific indications
of the neurologist. The patients who went
directly to the ward stayed in the recovery
room for 47±19 min. During this time used the
patient was monitored for sedation level and
blood losses, to optimize pain control, and to
gain ventilator weaning. The patients who
went into the ICU waited 28±8 min (Table 3).
Of the 4 patients discharged to the ICU, one
was already extubated in the OR. Another
patient was extubated after 20 min and the last
2 within 2 h of discharge from the OR. ICU stay
was no more than 24 h.
Average blood loss was 35±3 mL and no
treatment was needed; data are reported in
Table 4. Drain tubes were removed on Day 2.
Postoperative pain was treated by multimodal
analgesia with acetaminophen plus intrave-
nous (i.v.) morphine or acetaminophen plus
iv tramadol. The Visual Analogue Scale (VAS)
score was less than 4 at OR discharge and did
not increase at any time over the following
days. Length of hospital stay was 4.4±0.51
days. No patient was readmitted to the hospital
and no surgical complications were reported.
A single episode of seizure requiring treat-
ment was reported two days after surgery.

Discussion

In this paper we report the results we
obtained with 10 VATET in 10 pediatric
patients. Mainstay medical treatment of MG is
based on anticholinesterases and immunosup-
pressants. Thymectomy is suitable in patients
refractory to medical treatment or in patients
presenting moderate or severe forms of the
disease. Surgical removal of the thymus can be
useful to lower the level of antigenic response,
leading to remission in 30-70% of patients
after three years.2,8 The thoracoscopic
approach is also to be favored for esthetic pur-
poses, but especially for the reduction in the
use of chest drain tubes and painkillers, less
blood loss and shorter hospital stays.
Anesthesiological and intensive care man-
agement of myasthenic patients is well known,
as is the response of these to intravenous

Table 1. Myasthenia gravis Foundation of America clinical classification.

<table>
<thead>
<tr>
<th>Class</th>
<th>Clinical signs</th>
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<tbody>
<tr>
<td>Class I</td>
<td>Any ocular muscle weakness; may have weakness of eye closure. All other muscle strength is normal</td>
</tr>
<tr>
<td>Class II</td>
<td>Mild weakness affecting other than ocular muscles; may also have ocular muscle weakness of any severity</td>
</tr>
<tr>
<td>Class IIa</td>
<td>Predominantly affecting limb or axial muscles or both. May also have lesser involvement of oropharyngeal muscles</td>
</tr>
<tr>
<td>Class IIb</td>
<td>Predominantly affecting oropharyngeal or respiratory muscles or both. May also have lesser or equal involvement of limb, axial muscles, or both</td>
</tr>
<tr>
<td>Class III</td>
<td>Moderate weakness affecting other than ocular muscles; may also have ocular muscle weakness of any severity</td>
</tr>
<tr>
<td>Class IIIa</td>
<td>Predominantly affecting limb, axial muscles or both. May also have lesser involvement of oropharyngeal muscles</td>
</tr>
<tr>
<td>Class IIIb</td>
<td>Predominantly affecting oropharyngeal, respiratory muscles, or both. May also have lesser or equal involvement of limb, axial muscles, or both</td>
</tr>
<tr>
<td>Class IV</td>
<td>Severe weakness affecting other than ocular muscles; may also have ocular muscle weakness of any severity</td>
</tr>
<tr>
<td>Class IVa</td>
<td>Predominantly affecting limb, axial muscles, or both. May also have lesser involvement of oropharyngeal muscles</td>
</tr>
<tr>
<td>Class IVb</td>
<td>Predominantly affecting oropharyngeal, respiratory muscles, or both. May also have lesser or equal involvement of limb, axial muscles, or both</td>
</tr>
<tr>
<td>Class V</td>
<td>Defined by intubation, with or without mechanical ventilation, except when employed during routine postoperative management</td>
</tr>
</tbody>
</table>

The use of a feeding tube without intubation places the patient in class IVb.
anesthetics and gases.1

Both volatile and intravenous anesthetics are used during induction or maintenance of anesthesia; these do not affect weaning, extubation, or the postoperative (neither early or delayed) period.9

Paralyzing agents are not totally contraindicated, especially if these are short-lasting agents used with neuromuscular monitoring devices and new reversal drugs.6 This and the introduction of mini-invasive techniques have made it possible to markedly reduce the risk of perioperative complications in thoracic surgery, as well as the duration of mechanical ventilation and the length of stay in the ICU, improving outcome. This can also be seen in pediatric patients for whom the poor experience available in specifically designed clinical studies often induced the anesthetist to follow procedures normal for adult patients.

Clinical evaluation and neurological health are to be considered in the choice of postoperative ICU or ward discharge.

Pulmonary function evaluation is mandatory in the stratification of risk and in case of postoperative complexity.1 Anticholinesterase medications and corticosteroids are routinely stopped the day before surgery and are restored in the early postoperative phase. Premedication drugs (in our clinical practice, benzodiazepines) are not administered to avoid any additional risk of respiratory depression. We did not find any connection between the use of paralyzing drugs, delay in ventilator weaning/extubation time and discharge to ICU or ward.

The main cause of ICU discharge appeared to be preoperative borderline neurological status; certainly, early identification is made possible by interaction between the surgeon, the anesthetist and the neurologist.

Early extubation and discharge can be achieved in the majority of pediatric patients on completion of the surgical procedure. Only 40% of pediatric patients needed to be transferred to the ICU; all of them were in spontaneous ventilation and some of them were without the endotracheal tube. No delay in transfer from the ward to the ICU was required. Most of the conditions of discharge seemed to be inadequate and a post anesthesia care unit (PACU) appeared to be the ideal solution as an intermediate point of care between ward and ICU.

The ideal condition for choosing to transfer to the ward is also adequate analgesia, obtained with iv non-steroidal anti-inflammatory drugs (NSAID) and opiates. Paralyzing drugs seemed to be safe, thanks to the availability of monitoring of neuromuscular function and reversal drugs such as sugammadex.

The single case needing a neuromuscular paralyzing agent was later transferred onto the ward. Availability of new drugs, of reversal drugs, the current practice of mini-invasive surgical techniques, and the availability of PACU are the keys to safety and successful prognosis of patients affected by MG who undergo thymectomy.

### References