



SUBXIPHOID UNIORTAL VATS THYMECTOMY IN CASES OF MYASTHENIA GRAVIS:SINGLE CENTRE EXPERIENCE

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ABSTRACT **Background:** Thymectomy has been identified as Constructive and active strategy for patients with Myasthenia gravis and thymic masses which have been done classically by Open Sternotomy technique and nowadays new Minimally invasive approaches have also been introduced. **Method :** In this paper , Subxiphoid Uniportal VATS(Video Assisted Thoracoscopic Surgery) Thymectomy through a single port technique by utilizing the Subxiphoid approach has been discussed. I have improvised my technique pertaining to the requirements and better recovery and better post operative outcome of the patient .This procedure is indicated for all anterior mediastinal masses and maybe extended to lung cancer.The patient was placed in supine position instead of classical lithotomy position .Carbon dioxide insufflation was not used which led to faster recovery after the surgery.After dissection and resection of thymus Bilateral pleural drain were placed which was removed usually on Post Operative day 4 or 5 and patients were discharged afterwards. **Results:** Several benefits of this approach were observed and documented including reduced postoperative pain, Early extubation, better post operative outcome, and better dissection. **Conclusion:** In the near future ,the Subxiphoid approach has the potential to become GOLD STANDARD for Thymectomy and various other conditions

KEYWORDS : VATS(Video Assisted Thoracoscopic Surgery) ,Thymectomy, Subxiphoid

INTRODUCTION

Myasthenia gravis

Myasthenia gravis is a disorder of neuromuscular Junction caused due to auto antibodies targeting the junction and producing symptoms such as muscle weakness and fatigue which often oscillates during the course of the day. Auto Antibodies act against varied structures of Neuro Muscular Junction such as Acetylcholine receptor, Muscle specific tyrosine kinase, Low density lipoprotein receptor related protein 4, Neural Agrin,Nestrin 1 receptor CASPR 2 and Striated Muscle Antibody/titin.^[1]

Clinically it can be classified as Sero positive Myasthenia gravis where Anti acetylcholine receptor antibody are present which are most specific or Sero negative Myasthenia gravis where Anti musk antibodies ,Anti LRP -4 antibody . Anti Nestrin 1 receptor antibody,Anti CASPR 2 and Anti Striated Muscle Antibody/titin may be found.^[1]

Presentation of Myasthenia gravis in the OPD can usually be seen in either with age group 15 to 30 years or 52 to 65 years which can have gender prediction also.^[1]

In patients of earlier age group females are more commonly affected than men and can often be associated with other conditions such Addison's disease, Hashimoto thyroiditis, Vitiligo,Pernicious anaemia ,Type 1 DM and such patients often requires multi-disciplinary approach in management.^[2]

Early symptoms of Myasthenia Gravis are often limited to ocular muscles which gradually becomes a Generalized disease where proximal muscles of lower limb are frequently involved with delayed involvement of respiratory muscles.^[2]

However, patient can also present with ocular symptoms only being referred to as Ocular Myasthenia which can be Sero positive in 50% of cases but Anti Musk Ab are usually found. Ocular Myasthenia Patient often presents to OPD with severe buccal, facial, oropharyngeal involvement and Tongue Fasciculations.^[3]

Patients of Myasthenia gravis can often present to Emergency with symptoms such as Diaphoresis, Bradycardia, Bronchial secretions,Emesis, Lacrimation, Loose stools which can be due to Myasthenia crisis and can be managed by Intravenous immunoglobulin or plasma exchange therapy.^[1]

However apart from MG conditions such as chronic progressive external ophthalmoplegia and Lambert Eaten Myasthenic syndrome should also be considered as Myasthenia Gravis can often be associated with paraneoplastic syndrome.^[1]

Infants born to affected mother also show symptoms of the disease but recovers spontaneously by the age of two months coinciding with the disappearance of maternal antibodies suggesting the fact that neonatal myasthenia may be the autoantibody passively acquired from the mother.[19]

Clinical Grading of Myasthenia Gravis^[18]

MGFA Clinical Classification

Class I: Any ocular muscle weakness; may have weakness of eye closure. All other muscle strength is normal.

Class II: Mild weakness affecting muscles other than ocular muscles; may also have ocular muscle weakness of any severity.

A. IIa. Predominantly affecting limb, axial muscles, or both. May also have lesser involvement of oropharyngeal muscles.

B. IIb. Predominantly affecting oropharyngeal, respiratory muscles, or both. May also have lesser or equal involvement of limb, axial muscles, or both.

Class III: Moderate weakness affecting muscles other than ocular muscles; may also have ocular muscle weakness of any severity.

A. IIIa. Predominantly affecting limb, axial muscles, or both. May also have lesser involvement of oropharyngeal muscles.

B. IIIb. Predominantly affecting oropharyngeal, respiratory

muscles, or both. May also have lesser or equal involvement of limb, axial muscles, or both.

Class IV: Severe weakness affecting muscles other than ocular muscles; may also have ocular muscle weakness of any severity.

A. IVa. Predominantly affecting limb, axial muscles, or both. May also have lesser involvement of oropharyngeal muscles.

B. IVb. Predominantly affecting oropharyngeal, respiratory muscles, or both. May also have lesser or equal involvement of limb, axial muscles, or both.

Class V: Defined as intubation, with or without mechanical ventilation, except when employed during routine postoperative management. The use of a feeding tube without intubation places the patient in class IVb.

Thymoma

Thymus is a central organ for immunological self-tolerance and its functional disintegration often leads to assault on Acetylcholine receptor leading to Myasthenia gravis.^[20]

Most Thymic tumours in patients with Myasthenia gravis are Benign, Well differentiated and Encapsulated and can be removed completely at surgery.^[20]

Patients with thymoma have more severe disease, higher levels of acetylcholine Receptor Antibodies and more severe EMG abnormalities in Myasthenia Gravis.^[4]

Thymoma can have varied presentations either local symptoms due to Mass Effect causing symptoms like Pain in chest, blood in vomiting, Cough, Superior Vena Cava syndrome and Systemic syndromes caused by Immunologic mechanisms with Myasthenia Gravis being the most common.^[4]

Thymoma associated MG shows no gender bias, is rare in pediatric age group and peaks around 50 years of age.^[5]

In Thymoma, multiple antibodies are formed which are meant to attack on Thymus but due to structural similarity in Epitope, they are Misdirected towards Neuromuscular Junction leading to Myasthenia Gravis.^[6]

Thymectomy has been identified as Constructive and active strategy for patients with Myasthenia gravis and thymic masses which have been done classically by Open Sternotomy technique. But nowadays new Minimally invasive approaches have also been introduced.^[7]

Indications of Thymectomy.^[8]

1. Thymoma.
2. Good's syndrome
3. Acetylcholine receptor antibody positive.
4. Generalized Myasthenia gravis = 18-65 age group.
5. Bulbar involvement

Masaoka Staging System^[9]

- Stage I – The tumor has not spread into the outer layer of the thymus.
- Stage IIA – The tumor is starting to grow into the outer layer of the thymus or the nearby tissues and membranes.
- Stage IIB – The tumor has already grown into the outer layer of the thymus or the nearby tissues and membranes.
- Stage III – The tumor has started to grow into nearby tissues and organs.
- Stage IVA – The tumor has grown extensively into nearby tissues and membranes.
- Stage IVB – The tumor has spread to distant organs elsewhere in the body

TNM Staging of Thymoma^[10]

- TX: The primary tumor cannot be evaluated.
- T0 (T plus zero): No evidence of a primary tumor.
- T1: The tumor is located only in the thymus or has grown into the nearby fatty tissues.

- T1a: The tumor has spread into fat surrounding the thymus or
- T1b: The tumor has grown into the lining of the lung next to the tumor (called mediastinal pleura).

T2: The tumor has grown into the nearby fatty tissue and into the sac around the heart, called pericardium.

T3: The tumor has spread to nearby tissues or organs, including the lungs, the blood vessels carrying blood into or out of the lungs, or the phrenic nerve, which controls breathing.

T4: The tumor has spread to nearby tissues or organs, including the windpipe, esophagus, or the blood vessels pumping blood away from the heart.

Node (N)

NX: The regional lymph nodes cannot be evaluated.

N0: The tumor has not spread into lymph nodes

N1: The tumor may have spread to nearby lymph nodes.

N2: The tumor has spread to lymph nodes deep in the chest cavity or neck.

Metastasis (M)

M0 (M plus zero): The disease has not metastasized.

M1: The tumor has spread to other organs near the thymus, such as the lung and blood vessels.

M1a: The tumor has spread to the lining of the lung, called the pleura, or lining of the heart, called the pericardium

M1b: The tumor may have spread to the lining of the lung or the heart

Thymectomy

The main approach to Thymectomy conventionally has been through Median Sternotomy and however problems with Sternotomy include

- 1) Sternal dehiscence
- 2) Direct injury to Heart, lungs and Innominate vein
- 3) Infections after Sternotomy can contaminate middle Mediastinum with Skin flora and can cause Deep Sternal Wound Infection (DSWI) which still remains a difficult challenge for cardiac surgeons.^[11]

Minimally invasive approach.

The inevitable development of Modern technology has deeply influenced our approach to neoplastic and nonneoplastic Thoracic diseases.^[12]

Minimally invasive technique minimises Surgical trauma, shorten operative time and post operative stay improve outcome and better cosmesis.^[12]

We gradually Left Sternotomy, cervicostomy with Sternal split and adopted Minimally Invasive techniques which have tremendously improved the approach and outcome to mediastinum surgery.^[12]

Minimally Invasive Technique such as VATS can be done by multiple approaches such as Subxiphoid approach, Unilateral approach (right or left) and Bilateral approach.^[11]

Minimally Invasive Approach includes any approach as long as no sternotomy or thoracotomy with rib spreading is involved.^[15]

Advantage of Right-side approach includes Obvious Anatomical landmark and more easier to dissect the confluence of Innominate vein. But it is hard to see Left Phrenic nerve.^[13]

Advantage of left side approach includes easier dissection to the left side Anterior Mediastinal tumour but it is hard to dissect the Confluence of Innominate vein.^[14]

In bilateral approach complete resection is possible but involves

multiple incisions leading to poor cosmetic outcome and more post operative pain.^[15]

The technical considerations of Uniportal Subxiphoid approach would be discussed further in the article

Anatomy of Subxiphoid area and Mediastinum

Subxiphoid area is formed by the attachment of Linea Alba which is the continuation of the Rectus sheath. Once the Linea Alba is split and space is created following Anatomy is observed after inserting the scope.^[16]

Mediastinum is a broad Central partition that separates the two laterally placed Pleural cavity extending from Sternum to bodies of vertebra and superior thoracic aperture to diaphragm which can be divided into Superior mediastinum and Inferior mediastinum which can be further partitioned into Anterior, Middle and Posterior Mediastinum by the pericardial sac.^[16]

Anterior mediastinum contains loose connective tissue, mediastinal fat and body of thymus.^[17]

Thymus is Involved in the early development of the immune system; the thymus is a large structure in the child, begins to atrophy after puberty, and shows considerable size variation in the adult. In the elderly adult, it is barely identifiable as an organ, consisting mostly of fatty tissue that is some times arranged as two lobulated fatty structures.

From the anatomical Point of view, the thymus has 2 cervical poles in the neck and 2 poles in the mediastinum broadly extending along the pericardium and in Anterior costophrenic recesses.^[12]

Additional ectopic thymic foci have been found in the Pretracheal, Subcarinal and Anterior mediastinal fat, from the level of the thyroid to the diaphragm, in the aortopulmonary window, as well as from beyond each phrenic nerve, bilaterally.^[12]

An extended thymectomy has been described as the En bloc surgical removal of as much thymic tissue as possible, with surrounding mediastinal fat, from the cervical region to the diaphragm, also extending laterally to both phrenic nerves, also including the aortopulmonary window^[12]

Arteries to the thymus consist of small branches originating from the internal thoracic arteries.^[16]

Venous drainage is usually into the left brachiocephalic vein and possibly into the internal thoracic veins.^[16]

Lymphatic drainage returns to multiple groups of nodes at one or more of the following locations that is along the internal thoracic arteries (parasternal), at the tracheal bifurcation (tracheobronchial), root of the neck.^[16]

Surgery through Subxiphoid Approach.

Anaesthesia

Double-lumen endotracheal tube is used during anaesthesia. Intensive arterial monitoring is required because of the possibility of cardiac compromise during surgery with Left radial arterial line and catheterize Right Internal Jugular Vein Foley's catheterization is also being done .

Position.

Supine with arms by the side.

Surgical instruments

- (1) 30° thoracoscope (10 mm)
- (2) Grasper (5 mm), as long as possible (>30 cm):
- (3) Energy device: Harmonic scalpel, and a monopolar long tip cautery.
- (4) Vinyl bag medium sized.
- (5) Gonzalez Rivas dissector.
- (6) Soft tissue retractor.

Incision and port placement.

Incision.

A skin incision is made 4–5 cm Vertically at a single thumb's width down from the xiphoid process.

Port placement.

Uniportal instrumentation is done.

After finding the xiphoid process, the rectus muscle is transected about 1 cm to both sides and further dissection is performed 360° under the xiphoid process .

Using the index finger as in sternotomy, sufficient additional dissection is performed under the sternum and a soft tissue retractor is applied.

Thymectomy.

After sufficient space is available, the thymic tissue is dropped from the sternum. The boundary between the 2 sides is formed by the mammary vein, and dissection proceeds upward, closely adhering to the sternum. The mediastinal pleura is opened by reaching as far as possible where the mammary vein on both sides inserts into the innominate veins. After confirming that there is no bleeding, the thymic and pericardial fat is again dissected, as well as pleural fat from the inlet of the incision.^[17]

Thymoma should be resected by “No touch technique” to avoid rupture of the capsule and the risk of pleural dissemination.^[5]

During dissection, it is necessary to check the course of both phrenic nerves.^[17]

Bilateral Pleural Drain were placed

Post Operative Care

Daily Serial Chest X rays.

6 hourly Serial ABGs. on day 1 with removal of Arterial line and Foley's Catheter on the same day.

Early ambulation

Early initiation of Spirometry.

Decision to remove Bilateral pleural drain can be taken when Daily output is less than 20ml per day without any significant X ray changes. Although Sub-Xiphoid approach is having less post-operative pain which is better tolerated by the patient, Adequate Analgesia should be given to the patient.



DISCUSSION

ADVANTAGES OF SUBXIPHOID APPROACH

1) Patient's Perspective

- Reduced incision size
- Reduced Post operative pain
- Faster Recovery after the surgery
- Early extubation
- Better Cosmesis
- Decreased drainage as no bone is cut during the surgery
- Early discharge from the Hospital

2) Surgeon's Perspective

- Better dissection
- Better visualization of the anatomy
- Completeness of the dissection
- Lesser Chances of Phrenic Nerve injury
- Lesser adhesion and hence Re-operations or other operations can be performed with more ease.
- Lesser requirement of Anaesthesia and hence better post operative recovery.
- Rarely, CO₂ insufflation is being done at some centre to create artificial pneumothorax before surgery but due to use of Double Lumen EndoTracheal Tube with single lung ventilation, we have avoided Co₂ insufflation and hence there are reduced chances of Pulmonary Embolism and Co₂ retention.

DISADVANTAGES OF SUB-XIPHOID APPROACH. ^[17]

1) As this is a new approach and not practised everywhere, it requires experience on the part of operating cardiothoracic surgeon to be comfortable with all the aspects of surgery.

2) As it is an emerging technique, technological aspects of the surgery is not only dependent on the operating cardiac surgeon but on every single individual involved in the operating team who must be adequately trained to manage any Mis-happening or any other issues.

3) Although the operating time is less due to Direct Target Approach, WORKING OT TIME (Time period for which OT remains Functional apart from operative time) due to extensive logistical set ups before and after the surgery increases requiring more trained OT Staff.

CONCLUSION ^[17]

In the near future, the Subxiphoid approach has the potential to become GOLD STANDARD for Thymectomy and can also be applied to bilateral pneumothorax, bilateral pulmonary metastasectomy and simple lobectomy for both upper lobes and right middle lobe.

However proper case selection is necessary as when there can be multiple conditions in which we may encounter difficulties such as previous sternotomy scar or any previous surgery, or when there is advanced stage of thymoma and infiltration to the surrounding vascular structures is suspected.

In some cases Conversion to open approach is also required as there can be encasement of all the surrounding vascular structures when thoracoscope is inserted or when proper anatomy cannot be defined. This is an emerging technique but its accessibility can also be an issue of debate as this technique is still not being performed at every CardioThoracic centre.

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CONFLICT OF INTEREST

No conflict of interest relevant to this manuscript

ETHICAL STATEMENT

Not applicable

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