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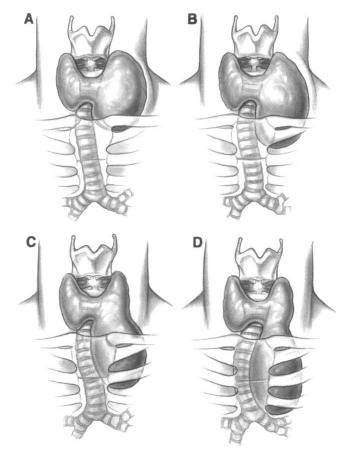
#### **Introduction:**

Thyroidectomy for a substernal goiter is typically a skill-demanding and complex procedure, yet very rewarding. The intricate neck anatomy is usually distorted in these cases, and often in an unpredictable pattern. Goiter size, shape, vascularity, distortion of anatomy, substernal extension, and restrictions imposed by the bones of the thoracic inlet can make thyroidectomy challenging, as well as recurrent laryngeal nerve (RLN) and parathyroid gland identification and preservation extremely difficult. That is why Halsted had said, "The extirpation of the thyroid gland for goiter better typifies perhaps than other operations, the supreme triumph of the surgeon's art."

This chapter is aimed at reviewing the patterns of anatomical distortion presented by substernal goiter, key points of preoperative evaluation, and treatment options with an emphasis on surgical approach.

#### **Definition and Classification**

There have been numerous definitions and classification schemes proposed for substernal goiter. They largely encompass the same definition; a goiter that is associated with a sub clavicular or substernal extension and therefore the thoracic component requires mediastinal dissection to facilitate extraction of the gland.



Classification of substernal goiters. (A) Grade I: <25% substernal; (B) Grade II: 26% to 50% substernal; (C) Grade III: 51% to 75% substernal; (D) Grade IV: >75%. Adapted with permission from Cohen et al. Operative Techniques in Otolaryngology-Head and Neck Surgery, vol 5, No 2 (June), 1994: pp 118-125

Moreover, a classification system is most useful when it incorporates the features of the substernal goiter that must be appreciated preoperatively for safe extraction. Thus, axial crosssectional imaging to differentiate between the various subtypes is necessary. We emphasize that IV contrast should be avoided in these cases, as the iodinated contrast can lead to acute goiter enlargement and worsen compressive symptoms, necessitating at times airway protection and urgent surgical intervention.



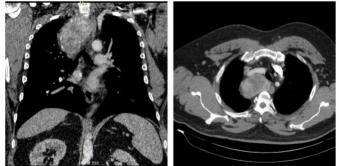
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| Туре | Location  | Anatomy   | Approach / Comment  |
|------|---|---|---|
| I    | Anterior mediastinum                              | Anterior to great vessels<br>(i.e., brachiocephalic and<br>subclavian vessels),<br>trachea, RLN | Transcervical (sternotomy, only<br>if intrathoracic goiter diameter ><br>thoracic inlet diameter) |
| п    | Posterior mediastinum                             | Posterior to great vessels,<br>trachea, RLN   | As above, also consider<br>sternotomy or right<br>posterolateral thoracotomy if                   |
| -IIA | Ipsilateral extension                             |   | - type IIB  |
| -IIB | Contralateral extension                           |   | -   |
| -B1  | Extension posterior to both trachea and esophagus |   | _   |
| -B2  | Extension between trachea and esophagus           |   | -   |
| ш    | Isolated mediastinal goiter                       | No connection to<br>orthotopic gland; may<br>have mediastinal blood<br>supply                   | Transcervical or sternotomy   |

•RLN: Recurrent laryngeal nerve

Substernal Goiter Classification - adapted from Randolph GW et al. Surgery of Cervical and Substernal Goiter. Surgery of the Thyroid and Parathyroid Glands  $-2^{nd}$  edition. Saunders 2013

When the thyroid gland extends into the anterior mediastinum (**Type I**), it descends anterior to the subclavian and innominate vessels and anterior to the RLN. The normal and orthotopic relationship of an anteriorly descended mediastinal goiter to the RLN is therefore preserved (i.e., the nerve is deep to the gland.)



Cross sectional computed tomography images of a patient with a large substernal goiter extending into the right mediastinum. The goiter descends anterior to the subclavian and innominate vessels and anterior to the RLN. (Substernal goiter type I)

When the substernal goiter extends into the posterior mediastinum (**Type II**), it assumes a space posterior to the trachea, pushing the trachea anteriorly and splaying the great vessels anteriorly. The mediastinal extension of the gland then rests in the space posterior to the innominate and subclavian vessels, carotid sheath, RLN, and inferior thyroid artery. Therefore, the normal relationship of the thyroid gland and the RLN is reversed. The RLN becomes ventral to the inferior component of the

thyroid and, if not recognized early on, can be stretched, or inadvertently cut. The nerve can also be entrapped between components of the posterior mediastinal goiter. This increased rate of RLN injury well documented in series comparing is thyroidectomy using only a cervical approach to sternotomy with mediastinal dissection to aid cervical removal. In those small series, there is a significantly decreased rate of RLN injury in patients who underwent sternotomy for mediastinal dissection.

Posterior mediastinal goiters can come to rest in a space bounded inferiorly by the azygous vein, posteriorly by the vertebral column, laterally by the first rib, medially by the trachea and esophagus, and anteriorly by the carotid sheath, subclavian and innominate vessels, superior vena cava, and phrenic and RLNs. Therefore, it is important to be familiar with the regional anatomy of the lower neck and upper mediastinum.

Posterior mediastinal goiters can occur on the same side as the enlarged lobe (**Type IIA**) or may come to rest through a retro-tracheal extension in the contralateral thorax (Type IIB.) Extension to the right mediastinum is more commonly seen because of the aortic arch and associated branch vessels blocking the left posterior mediastinal descent pathway. Contralateral posterior mediastinal extension may occur either behind the trachea and esophagus (Type IIB1) or between the trachea and esophagus (Type IIB2). Generally, the right mediastinal caudal extension is limited at the level of the azygous arch.

Although rare, thyroid masses within the mediastinum may exist without connection to the normal cervical thyroid gland, known as an ectopic thyroid. Isolated mediastinal goiters are important to recognize because unlike all other types of substernal goiters, blood supply of the goiter may be through purely mediastinal arteries (i.e., aorta, subclavian, internal mammary, thyrocervical, and innominate) and veins. This is extremely important in planning their surgical resection. Also worth noting is that it is not uncommon for a goiter to be dumbbell shaped, when the substernal component is attached to the cervical component through a narrow thin band of



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thyroid parenchyma. Noting an ectopic thyroid goiter or a dumbbell shaped thyroid goiter preoperatively is crucial as complete resection is very difficult to achieve through a cervical approach only, even if the substernal component is not large or does not extend significantly into the mediastinum.

## Workup

In the workup of patients presenting with a substernal goiter, the surgeon needs to systematically investigate all the following issues preoperatively: 1. Existence of compressive symptoms such as:

- a. Voice change or voice fatigue which could hint at an RLN already under excessive tension from displacement
- b. Dysphagia and regurgitation which could indicate significant mass effect on the esophagus, and the greater risk of an aspiration event upon induction of general anesthesia
- c. Airway compression, especially when the patient lies flat and is fully relaxed upon induction (i.e., loses most cervical muscle tone and the full weight of the goiter pushes down or further deviates the trachea). It is also important to realize that chronic mass effect of the goiter on the trachea results in tracheomalacia, and subsequently can leave the patient with a high risk of airway obstruction even post thyroidectomy.
- 2. Risk of malignancy, especially if the portion of the thyroid that contains the suspected malignancy is substernal and not accessible for percutaneous needle biopsies to establish the diagnosis.
- 3. Presence of hyperthyroidism, which would require medical therapy prior to thyroidectomy to achieve a euthyroid state and minimize the risk of a thyroid storm intraoperatively. Also, the risk of bleeding during thyroidectomy for a toxic thyroid goiter is slightly greater as they are commonly more vascularized than nontoxic goiters.
- 4. Integrity of the RLN should be assessed either through ultrasound evaluation of the mobility

and symmetry of the vocal cords (which can be challenging in patients with large goiters) or via direct visualization with nasolaryngeal fiberoptic endoscopy.

5. Subtype of mediastinal extension (Table, above) and preoperative consultation with potential surgical planning by a thoracic surgeon, if available. The majority of substernal goiters will be resected through cervical approach. But there are a few characteristics that increase the risk for an extra-cervical approach: Malignant histology, Extension below the aortic arch's convexity in the cranio-caudal plane and posterior extension to trachea in the anteroposterior plane.

It is important, in limited-resource countries, that the surgeon be as independent as he/she can at imaging interpretation.



Goiter physical examination: lateral and anterior view



Find a "sulcus" between thyroid and sternum's manubrium, with mobile thyroid gland are a sign that there are minor or no



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mediastinal extension. Thus, a cervical approach is more likely to succeed.



As seen here, a widened mediastinum (on the right) and deviation of the trachea (towards the left) are indications that the airway is compromised and intubation may be more difficult.

## Indications

It is generally agreed that thyroxine suppression therapy and radioactive iodine ablation are not acceptable interventions, and that surgery is the gold standard for substernal goiter management. Overall, these are the major indications for substernal thyroidectomy:

- 1. Suspicion for or confirmed thyroid cancer
- 2. Compression of adjacent organs
- 3. Toxic multinodular goiter or toxic adenoma or thyrotoxicosis
- 4. Cosmetic reasons

Most surgeons would favor thyroidectomy even if asymptomatic, as substernal goiters will continue to grow over time and become more complex to resect when larger in size or extend further into the mediastinum. Moreover, the thoracic component of a substernal goiter poses difficulty for ongoing clinical examination or fine needle aspiration, as mentioned above.

Surgery usually comprises two different approaches; one is completely transcervical, in which only a cervicotomy is performed, and the other requires an additional extracervical approach (e.g., thoracoscopically or via sternotomy.) Several different surgical techniques have been described; however, no clearly defined consensus exists on the indications for either approach to substernal thyroid goiter excision.



A retrosternal goiter delivered through a trans-cervical approach. The dissection of the inferior part of the enlarged lobe was done with the surgeon's finger, in the capsular plane of the thyroid.



The completely removed specimen. The left lobe is 11cm in craniocaudal length, and more than half of that was inside the thoracic cavity, yet it could be removed through a transcervical approach.

## Steps:

#### Intubation

Intubation is usually uneventful in patients with substernal goiters, but sometimes can be



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difficult, or even life-threatening, requiring an emergent and very challenging tracheotomy due to the overlying engorged goiter. A small portion of patients may also have evidence of substantial laryngeal deviation or vocal cord paralysis at intubation. A large substernal goiter can compress the venous drainage of the neck causing venous congestion that results in pharyngeal, laryngeal, and supraglottic soft tissue edema. Therefore, the surgeon and anesthesiologist should review all the preoperative data including cross sectional imaging and preoperative laryngoscopy and, examine the patient together before induction. The method of intubation. size of endotracheal tube. and contingency plans can be discussed and decided upon through these discussions.

Typically, a straightforward induction with transoral intubation can be performed. Laryngeal deviation does not generally pose difficulty and a reasonably sized endotracheal tube can be used despite the extrinsic tracheal compression. A safe alternative method is an awake fiberoptic transoral or transnasal intubation with the patient in a full sitting position. Full knowledge and access to the maneuvers and instruments that can be employed for airway protection and better visualization of the vocal cords should be readily available. Newer video laryngoscopes are also an excellent adjunct for intubation in such patients. Ultimately, to decrease the risk of laryngeal edema from intubation, it is best to intubate once correctly, as studies have shown that the larynx is easily made edematous with multiple unsuccessful intubation attempts. A useful maneuver, especially during awake intubations, is to grab the tongue with a dry gauze and pull it forward as much as the patient can tolerate. This will anteriorly displace the swollen laryngeal and supraglottic apparatus and improve cord visualization. "Armored" endotracheal tubes can be used for patients with significant tracheal compression distally. See also the chapter Airway Management and Endotracheal Intubation.



Fiberoptic transoral intubation. The epiglottis is seen on the monitor. Note that the endotracheal tube is already on the fiberoptic scope: once the scope enters the trachea, this can be slid along the scope into position.

## Extent of Surgery

The of extent surgery (i.e. hemithyroidectomy or total thyroidectomy) should be tailored to allow a favorable balance between the risk of operative complications and disease progression or recurrence. Complication rates must be kept extremely low in the setting of benign disease. Therefore, the minimum procedure should be a hemithyroidectomy, reserving bilateral surgery for significant bilateral goiter, malignant disease, or toxic multinodular goiter/ diffuse hyperthyroidism. Sometimes, when an ipsilateral nerve injury or an ipsilateral loss of nerve signal during intraoperative nerve monitoring occurs, the surgeon must change plans intraoperatively and possibly defer resecting the contralateral lobe. This helps avoid a potentially disastrous case of bilateral RLN injury requiring tracheostomy.

## **Positioning**

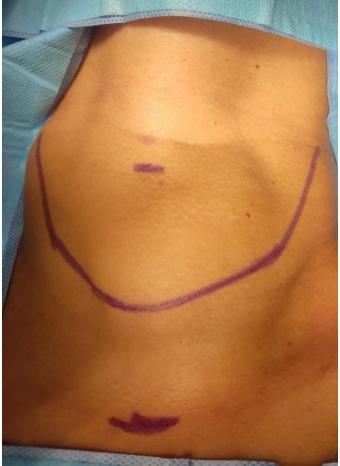
Great emphasis should be placed on proper positioning of the patient on the operating room table. A shoulder roll should be placed under the patient's shoulder blades as to extend the neck safely as much as possible with the head well supported. Additional extension may be obtained by placing the patient in a beach-chair mode position.



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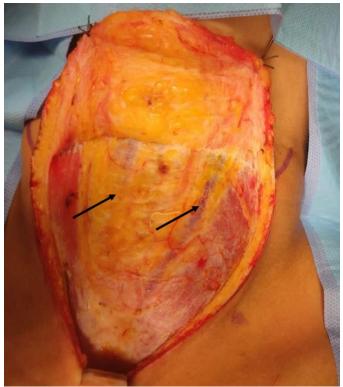
## Incision and Subcutaneous Flaps

A generous collar incision is usually needed. Minimal access approaches are not appropriate for substernal goiter extrication. The traditional Kocher incision, extending to the lateral edge of the sternocleidomastoid (SCM) muscle, may be required for large bilateral goiters.



A simple transverse collar incision is often sufficient. For very large glands, such as this one, the incision should be extended upwards along the anterior border of the sternocleidomastoid. Extending your incision further posteriorly does not help as much to improve access, and gives the appearance of a "decapitation."

A subplatysmal upper flap is developed all the way to at least the level of the cricoid cartilage; consider going to the level of the hyoid bone in especially large goiters. A generous lower flap is typically not necessary. Flaps can be sutured in place, or a self-retaining retractor can be used.



The same incision, now with the flap elevated in the subplatysmal plane. Note the anterior jugular veins (Black arrows.) You will be dissecting along these, so proper technique is important to avoid troublesome bleeding." Note also that this vein should be ligated if the "strap" muscles are divided, as described below.

## "Strap" Muscles

Routine transverse sectioning of the strap muscles (sternohyoid and sternothyroid) during thyroidectomy for substernal goiter is not needed. Preserving the strap muscles allows preservation of the planes and anatomic organization of the central compartment and neck base that can be substantially altered by the goitrous change. However, if there is any question that strap division would improve mobilization, it should be done. Transverse sectioning of the strap muscles should occur at the top third of the muscle as the nerve innervation comes in caudal to cranial. The strap muscles can then be sutured back together at the completion of surgery with little adverse effect. The anterior jugular vein, running along the anterior surface of these muscles, may be sizeable in these patients and should be separately divided and ligated.



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## Early Division of the Isthmus

Early division of the thyroid at the isthmus, even in the setting of total thyroidectomy, facilitates better exposure, retraction and RLN identification in these cases. Dividing the isthmus early can allow for more medial mobilization of the thyroid lobe to facilitate exposure of the RLN as discussed later in this chapter. This might be more challenging when the most inferior aspect of the isthmus extends substernally along with the substernal component of the thyroid lobe. A key step to successfully perform this maneuver is to correctly identify the plane, using blunt dissection, above the cricothyroid muscle where its fibers meet along the cricoid cartilage and the isthmus. If the plane below the cricothyroid muscle is entered the dissection will be bloody and the risk of inadvertent penetration of the cricothyroid membrane or even trachea is more likely.

# Recurrent Laryngeal Nerve Identification and Preservation

The most important rule to follow is that no structure is cut until the RLN is identified visually and, if nerve monitoring used, electrically. A bloodless field is essential for identification and preservation of the RLN. When operating on large goiters, this requires an especially careful technique. Once the inferior and superior poles are dissected, the strap muscles are retracted laterally and the thyroid, along with the trachea, should be retracted as one unit medially. This opens the lateral thyroid region. As the thyroid is dissected and freed from its lateral cervical attachments, it is progressively pulled medially and the airway itself is to some degree displaced upwardly and rotated through this retraction. This will expose the nerve at the RLNinferior thyroid artery crossing junction. Gentle blunt dissection helps in this maneuver. In this approach, the nerve is not uncovered inferiorly at the thoracic inlet. This approach helps to preserve parathyroid vascular supply, especially for the inferior parathyroid. Care must also be taken on the right side to identify a nonrecurrent right RLN.

In certain cases of large goiters or in cases in which the tubercle of Zuckerkandl is well developed, the lateral thyroid region may not be adequately exposed to permit easy RLN identification. In such circumstances the nerve is best identified inferiorly. This involves identification of the RLN in the lateral aspect of the thoracic inlet on the right and in the paratracheal position at the thoracic inlet on the left. This relationship is shown in the photo below. Advantages of nerve identification at this location include the soft areolar bed in which the nerve lies in this region, allowing for atraumatic dissection. Another advantage of finding the nerve in the thoracic inlet is that the nerve here exists as one trunk before extra-laryngeal branching, which can occur above the inferior thyroid artery crossing point.

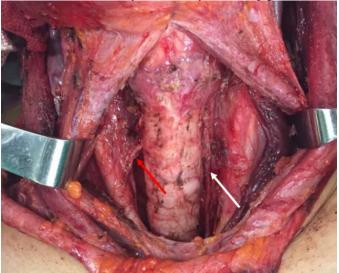


Photo after removal of the thyroid gland shows the course of the right and left recurrent laryngeal nerves. The right nerve (Red arrow) courses around the right subclavian artery, which is very close to the area being dissected. Therefore, the nerve must take a more medial-to-lateral course to reach the larynx. The left nerve (White arrow) courses around the aortic arch, which is farther down in the chest and closer to the midline. It will therefore be taking a direction that is more straight craniocaudal.

In some cases of large cervical or substernal goiters where the size and distortion of the anatomy make the lateral or inferior approach to identifying the RLN impractical, identifying the early nerve at the ligament of Berry-laryngeal entry point can be done. To facilitate this approach, early transection of the isthmus and initial dissection of the superior pole allow for lateral and caudal retraction of the superior pole to allow access to this segment of the nerve during the initial phases of lobectomy. The inferior



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cornu of the thyroid cartilage can be palpated to assist in nerve location in this area. The disadvantage of the RLN identification through this approach is that the dissection occurs at the ligament of Berry, which is fibrous and bleeds easily. Also, the nerve at this region may be branched. This approach is technically more challenging with large superior poles. The exposure of this superior pole region can be improved by division of the sternothyroid muscle, as described above.

#### Parathyroid Preservation

The distal inferior thyroid artery is divided after the RLN is identified and either before or after goiter delivery. The artery is ligated and divided directly on the thyroid capsule to reduce the risk of parathyroid ischemia. The superior parathyroid glands are more constant in position and are more frequently seen at thyroidectomy for goiter; therefore, they are more readily preserved. The inferior parathyroid gland is more variable in position and more likely to be significantly displaced by inferior pole goitrous change. Therefore, real emphasis during goiter surgery should be on superior parathyroid preservation. Inferiorly, strict adherence to capsular dissection to preserve displaced inferior parathyroid glands should be performed. Lastly, any resected thyroid specimen must be meticulously examined for capsular parathyroid glands before being sent to pathology. Any capsular parathyroid glands that are found should be dissected off, biopsied to confirm parathyroid tissue, and then auto-transplanted. Briefly, the parathyroid gland is cut into many tiny pieces with fine scissors or scalpel, and then placed within a surgically created "pocket" of adjacent muscle, usually the medial edge of the sternocleidomastoid. Recall that parathyroid glands may be found within folds and crevices of the goiter surface.

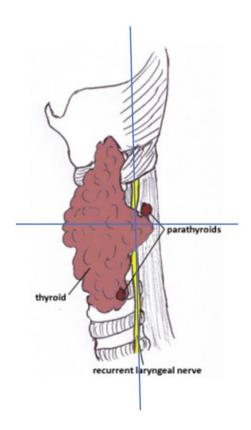


Diagram showing the normal location of the parathyroid glands relative to a vertical line drawn through the axis of the recurrent laryngeal nerve, and a horizontal line drawn through the axis of the tubercle of Zuckerkandl, the posterior-most extent of the thyroid gland. The superior parathyroids, needing to migrate less distance, are more consistent in their location. The inferior parathyroids, having migrated farther and possibly being displaced by the goiter, are more likely to be in an abnormal position. Source:

http://www.endocrinesurgery.net.au, used with permission.

## Delivery of the Goiter from the Mediastinum

After the RLN is identified and completely dissected away from the goiter, finger dissection in the capsular plane can allow for safe goiter delivery. The goiter is slowly mobilized upward out of the chest. Fascial band attachments as well as vascular pedicles are drawn up gently with the surgeon's finger are cauterized or clamped only after the RLN location is completely identified along its entire course. Nerve monitoring can also be extremely helpful in this part of the procedure. Cysts within the thyroid, if benign, can be decompressed intraoperatively with a large bore needle to decrease the gland's overall size.



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A variety of instruments have been used to facilitate substernal goiter delivery. A mediastinal goiter "spoon" and a foley catheter placed in the mediastinum have both been described to assist in the delivery of substernal goiter without sternum division. If all these maneuvers are not effective, partial or complete sternotomy, or a thoracoscopic approach may be needed. It is important that the thyroid surgeon consults with their thoracic surgical colleagues and arrange the surgical date when a thoracic surgeon is available.

## Pitfalls

- A complete preoperative workup is essential in treating patients with substernal goiters. Preoperative identification of substernal extension of the thyroid allows for appropriate preoperative imaging and surgical planning. Moreover, early identification of compressive symptoms or RLN dysfunction helps avoid perioperative complications. If CT scan is not available. careful physical examination supplemented by plain x-rays and ultrasound allows the surgeon to make a reasonable estimate of the extent of the substernal portion.
- An anesthetist should review all the preoperative data to include cross sectional imaging and preoperative laryngoscopy and, examine the patient before induction. This ensures that the proper intubation technique and perioperative care will be administered.
- Like routine thyroid surgery, the most important rule to follow is that no structure is cut until the RLN is identified visually and, if nerve monitoring used, electrically. A bloodless field is essential for identification and preservation of the RLN. The surgeon should be aware of the different techniques for RLN identification and preservation during these difficult cases.
- Parathyroid identification and preservation is as important as RLN identification in these cases. Real emphasis should be placed on superior parathyroid preservation. Inferiorly, strict adherence to capsular dissection to preserve displaced inferior parathyroid glands should be performed. Any resected thyroid specimen must

be meticulously examined for capsular parathyroid glands, which are then auto-transplanted.

• Preoperative surgical consultation with thoracic surgical colleagues, if available, is important and arrangements must be made so that proper intraoperative and postoperative care can be administered if the patient were to require sternotomy or thoracoscopy to assist with delivery of the substernal component. Further description of the sternotomy incision is in another Chapter of this Manual.

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