Substernal goiter: when is a sternotomy required?

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Abstract

**Background:** The presence of substernal goiter is, per se, an indication for surgical management. The aim of this study was to identify the preoperative predictors of a sternotomy in the management of substernal goiter in order to provide better preoperative planning and patient consent.

**Methods:** Between 2005 and 2008, 535 patients were referred to our clinic for thyroidectomy, 34 patients (6.3%) had substernal goiter and were included in this study.

**Results:** All substernal goiters were treated surgically, 31 (91%) by a cervical approach and 3 (9%) by full median sternotomy. All surgeries were successful, with no major postoperative complications. The minor postoperative complications transient hypocalcemia and transient paralysis of the recurrent laryngeal nerve occurred in 4 (11.7%) and 2 (5.8%) cases, respectively. The indication of the median sternotomy were as follows; extension of goiter below the aortic arch, tracheal deviation and, large thyroid tissue extending towards tracheal bifurcation and ectopic thyroid tissue in the mediastinum.

**Conclusions:** Substernal goiter, can be removed through a cervical incision, but on rare occasions a median sternotomy may be required.

**Key words:** sternotomy, substernal goiter, surgery, treatment
Introduction

Substernal goiter (SG) was first described by Haller in 1749 and first surgically removed by Klein in 1820 [1,2,3]. There is no uniform definition of substernal goiter [1,2,3]. However, various different criteria have been suggested by authors. These include a thyroid gland extending 3 cm below the sternal notch or extension of the gland below the fourth thoracic vertebra [4,5]. An extension of the thyroid gland below the thoracic inlet has been defined as substernal, retrosternal, intrathoracic or mediastinal goiter. Drawing upon the relationship of the intrathoracic extension of SG to the arcus aorta and the right atrium and findings from imaging methods, diagnostic classifications have been established that take into account the percentage of goitrous thyroid in the mediastinum [6,7]. Substernal goiters are common, with a reported incidence of 1-20% of all patients undergoing thyroidectomy [3,4,8-12]. Diagnosis of substernal goiter is most frequently made in the fifth or sixth decade of life, with a female/male rate of 4:1 [10,13,14]. The vast majority of SGs (85-90%) are located in the anterior mediastinum with the remainder (10-15%) located in the posterior mediastinum [1,9,15,16].

Substernal goiters show, in most cases, a slow-growing enlargement, which usually remain asymptomatic for many years; about 20-40% of substernal goiters are discovered as an incidental finding on a radiographic examination [2,10,17]. Patients with mediastinal goiter are rarely asymptomatic. The most common symptoms are related to compression of the airways and the oesophagus, and represented by dyspnea, choking, inability to sleep comfortably, dysphagia and hoarseness [2,11,13,17]. The diagnosis of substernal goiter is based upon clinical history, clinical examinations, and imaging findings. In the diagnostic management of mediastinal goiter, chest computed tomography (CT) was of the highest value. CT scanning is, at present, the most exhaustive examination for assessment of the extent of the goitre and compression effects on adjacent anatomical structures. A preoperative CT
scan should be routinely performed in every suspicion of a substernal goiter [11,18]. Magnetic resonance imaging (MRI) adds little additional information to that obtained with CT and is not routinely used [10].

Substernal goiter, must be removed surgically due to relation to compressive symptoms, potential airway compromise, and the possibility of an association with thyroid malignancy [16]. There is a general consensus that most can be successfully removed via a cervical approach and that thoracic access is rarely necessary [8,10,11]. Various factors have been reported to increase the likelihood of a median sternotomy being required. These factors include involvement of the posterior mediastinum, extension of the goiter to the aortic arch, recurrent goiter, superior vena cava obstruction, malignancy with local involvement, and emergent airway obstruction [5,9,11,15,16,18]. In addition, inability to palpate the lowermost extent of the gland also is considered to be an indication for median sternotomy. The incidence of sternotomy in substernal goiter is variable, ranging between 0-11% [2,8,10,11]. This wide range in incidence might be related to variation in the definition of substernal goiter.

In order to improve preoperative planning and patient consent, we aimed to identify the preoperative predictors of a sternotomy in the management of substernal goiter.

Materials and Methods

In this retrospective study, we evaluated the medical records of 34 patients diagnosed with substernal goiter out of a total of 535 thyroidectomies performed in our clinic between 2005 and 2008. The goiter was defined substernal when extending at least 3 cm below the thoracic inlet performed with hyperextension of the neck ascertained by preoperative imaging methods and confirmed intraoperatively. In all those patients with clinical suspicion of substernal extension of the goitre (lower pole of the cervical goitre not palpable) or with fi
ndings of tracheal deviation or a mediastinal mass upon chest X-rays and ultrasound findings, neck and chest CT scans were obtained to get an accurate delineation of the goiter size and its relation to the adjacent structures. Each patient was subjected to preoperative chest X-ray to check for tracheal deviation and any mediastinal mass. Furthermore, various imaging methods such as thyroid ultrasound, neck and thorax CT, and thyroid scintigraphy were employed, depending on the case. Preoperative CT scan analysis has focused on maximal substernal area of the goiter and its relation to the trachea, esophagus, and major vessels. The surgical technique, intraoperative findings, and postoperative course were recorded for all patients. This study was approved by our institutional ethics committee.

**Results**

Of the 535 patients who underwent thyroidectomy, 34 (6.3%) had SG. Of these, 21 (60%) were female with a mean age of 50 (24-68) and 13 (40%) were male with a mean age of 53 (18-76). The most common symptoms included a palpable cervical mass, dyspnea and dysphagia. The duration of these symptoms was 2 months to 3 years. Diagnoses were made by chest X-ray, thyroid ultrasonography, neck and thorax CT and MRI, and thyroid scintigraphy, in addition to clinical examination, and were confirmed intraoperatively (Figures 1, 2, 3). The substernal extension was well below the aortic knuckle at the tracheal bifurcation in three patients who required sternotomy. Three sternotomy patients had radiologic evidence of tracheal deviation, compression, or both (Figure 1,2,3). All the patients had a euthyroid preoperative status. Each was preoperatively examined by indirect laryngoscopy and none had vocal cord paralysis. Fine needle aspiration biopsy was used in one patient. No patient had a history of previous thyroid surgery. Of the 34 patients, 31 (91%) thyroidectomy was performed through a cervical incision, while in 3 patients (9%) a full median sternotomy had to be performed, in addition to the cervical incision. On operation, the substernal component was seen on the left in 21 (61.8%) patients, on the right in 9
(26.5%) patients, and on both sides in 3 (8.8%) patients and in one (2.9%) patients ectopic location. All the patients underwent bilateral total thyroidectomy. There was no operative mortality, blood transfusion requirement, need for tracheostomy, or wound infection. To prevent hematoma development, a meticulous surgical technique, good hemostasis, and negative pressure drainage for 48 h were used. Postoperative complications occurred in 6 (17.6%) patients, 4 (11.7%) of whom developed transient hypocalcemia and 2 (5.8%) transient recurrent laryngeal nerve (RLN) paralysis. The patients who developed transient hypocalcemia were treated with oral calcium. Following this treatment, their serum calcium levels reverted to normal and no other medications were needed. The patients who developed transient RLN paralysis received anti-edema treatment and their clinical condition improved after 7 d. The duration of hospital stay of the patients was 3 d on average. Those with transient hypocalcemia stayed for an additional 2 d and those with transient RLN paralysis had an additional stay of 5 d. Postoperative histopathologic examinations revealed malignancy in 7 (20.6%) of 34 patients. Of these 7 patients, it was found that 3 had papillary carcinoma, 3 had follicular carcinoma, and 1 had beta cell lymphoma, while in the remaining 27 patients (79.4%) a benign multinodular goiter was discovered. In one of 3 patients with follicular carcinoma had performed full median sternotomy. Patients were followed up for an average of 2 years (6 months to 40 months) and no recurrence was observed.

Discussion

Substernal goiters can cause respiratory distress, dysphagia, vascular compression, and even sudden death. They are not uncommon and carry a risk of malignancy between 3–21% [8,13]. Because there is no effective treatment other than surgery and because thyroidectomy is felt to be both safe and effective, the presence of substernal extension is an indication for surgery in an otherwise healthy patient, even in the absence of clinical symptoms [8]. Surgical removal of substernal goiter can be performed by a cervical approach.
in the majority of patients [8,10,11]. It has been reported that skilled surgeons, with good
thyroid surgery experience, need to perform an extra-cervical approach in 2-5% of
thyroidectomies for substernal goiters, but some authors have reported an incidence of
 sternotomy in 29% of patients [2,11]. In our study, sternotomy was required in three
patients (9%). This variability could be correlated with the lack of uniformity in definition of
substernal goiters [8,10]. Crile defined mediastinal goiters as those that extend to or inferior to
the aortic arch, whereas Lahey required that the greatest diameter of the goiter be inferior to
the thoracic inlet on chest X-ray. Others have used a more liberal definition of substernal,
including goiters with any degree of extension into the thoracic inlet [1,8,10]. Substernal
goiter was defined by de Souza and Smith as a goitre with a portion of its mass ≥ 50%
situated in the mediastinum [10]. More precise definitions of substernal goiter have been
suggested, namely: a goitre lying two fingerbreadths below the thoracic inlet with the patient
in a supine position, a goitre reaching the aortic arch, or the carina tracheae, a goitre with its
inferior pole passing through the cervico-thoracic isthmus below the subclavian vessels [10].

In our study, we considered as substernal those goitres showing an extension of at least
3 cm below the cervico-thoracic isthmus, at CT scan, performed with hyper-extended neck,
excluding from the study all the many other goiters showing a less mediastinal extension.

Mediastinal goiters can remain asymptomatic until compression of the structures located
in the thoracic inlet occurs. Life-threatening mechanical compression can occur because of the
limited space below the thoracic inlet. The most common symptoms are dyspnea, dysphagia,
cough and hoarseness. Some patients present with superior vena cava syndrome. In our one
patients had superior vena cava syndrome.

There is general consensus that the vast majority of substernal goiters can be
successfully extracted through a cervical approach. However, in patients in whom the gland
cannot be safely removed through a cervical incision, thyroidectomy may require a
sternotomy. In our series, SG could be removed, using a standard cervical approach, in 31/34 patients (91%), despite the large size and depth of the mediastinal extension of the goitres. However, a sternotomy was required in 3/34 patients (9%), due to extension of substernal goiter below the aortic arch in one patient, and a very large thyroid, reaching the main bronchial bifurcation in the other patients (Fig1,2,3).

Following preoperative imagining methods, 3 patients had to undergo full median sternotomy (Figure 1,2). One of these two patient’s goiter was extending below the aortic arch, sternotomy. These 2 of 3 patients had tracheal deviation and mediastinal mass in their chest X-ray and one patient’s thyroid was very large and extending towards tracheal bifurcation in neck and chest CT scan and MRI imaging. (Figure 1,2). Both of the patients had significant predictive factors of the need for sternotomy since the patients had significant tracheal deviation and the goiter was extending below the aortic arch. In addition, goiters larger than 10 cm were identified by preoperative imagining methods in these 2 patients, and this finding was also evaluated as a predictive factor of the need for sternotomy. (Figure 1,2). Besides, one of these two patients had vena cava superior syndrome. Vena cava superior syndrome is an indication for median sternotomy.

A mediastinal mass was identified in the chest X-ray of the 3rd sternotomy patient. A mediastinal mass of 15 cm was extending towards tracheal bifurcation in neck and chest CT scan and MRI imaging. Fine needle aspiration biopsy was performed and the mass was identified as thyroid tissue. Following thyroid scintigraphy, CT scan and MRI imaging, it’s observed that this mass was not connected to the cervical thyroid, but an ectopic goiter. (Figure 3). Because of its size and ectopic location, full median sternotomy with cervical incision was performed. Ectopic goiter is another preoperative predictive factor of sternotomy.
In CT scans of 3 patients, mediastinal goiter and evidence of adherence to the surrounding mediastinal tissues and great vessels are further strong predictive factors for median sternotomy.

For surgeons performing thyroidectomy, it is essential to pre-operatively identify patients requiring sternotomy, in order to plan the presence of a multi-disciplinary team, involving also the thoracic surgeon, when necessary, and to correctly inform the patient about the approach. Many attempts have been made to specifically define the factors increasing the likelihood of sternotomy, but a general consensus has still not been reached [10]. Solid criteria have not been established yet for selecting substernal goiter patients who would probably require a sternotomy.

Despite the widespread use of CT scans in the preoperative assessment of substernal goiters, the correlation between CT scan features and the surgical approach required has been poorly investigated to date. Furthermore, attempts to establish any significant correlation between the patients clinical features and the actual surgical procedure always proved fruitless. Casella et al. found that extension of the goiter below the level of the aortic arch appeared to be a significant predictive factor for the need for sternotomy [15]. Conversely, the lack of radiologic extension beyond the aortic arch predicted successful transcervical removal of mediastinal goiters without sternotomy [15]. At CT scan evaluation, suggesting that the need for a sternotomy is related more to anatomic location than to the actual size of the goiter.

Substernal extension of mediastinal goiters with respect to the trachea and major vessels did not appear to be, per se, a predicting factor for sternotomy; however, posterior mediastinum involvement by the gland was found as a further strong predictive factor for sternotomy need when present along with extension of the goiter below the aortic arch.

Several series have examined the factors that increase the possibility of sternotomy. Flati et al. stated that sternotomy is inevitable when a goiter is iceberg shaped and more than
70% of it resides within the mediastinum [5]. Some authors have listed revision surgery as a possible indication for sternotomy. De Perrot et al. felt that sternotomy should only be performed in cases of revision surgery, invasive cancer or ectopic goiter. Mussi believed that sternotomy should be employed when a goiter could not be extracted from the chest with ‘gentle maneuvers,’ as well in cases of all recurrent and aberrant goiters [19]. Sand et al. employ sternotomy when excessive traction is required during surgery, when the most inferior extent of the nodule cannot be palpated, in cases of revision surgery, in the setting of acute tracheal compression, severe venous obstruction, malignancy, and uncertain preoperative diagnosis [20]. Sancho felt that nodules that extended inferiorly to the level of the carina placed patients at high risk for sternotomy [21]. Randolph recommended sternotomy for malignant substernal nodules, posterior mediastinal goiter with contralateral extension, mediastinal goiters with mediastinal blood supply, goiters causing superior vena cava syndrome, revision cases, in the setting of difficult delivery from the chest, significant hemorrhage, and when the diameter of the mediastinal nodule significantly exceeds the diameter of the thoracic inlet [8].

One of the factors increased the likelihood of sternotomy is malignancy. The incidence of malignancy in retrosternal goiters has been reported between 3–21% [8,12,16,17]. Although fine needle aspiration biopsy is often impossible in nodules that are located completely within the mediastinum, advanced malignancy can often be suspected on the basis of preoperative MRI or CT scan. Thus, malignancy does not automatically mandate sternotomy, but thoracic access should be considered for malignant nodules that may be adherent to mediastinal structures or blood vessels and larger cancers whose transit through the thoracic inlet might result in tumor spillage. Nevertheless, Rugiu and Piemonte still consider malignant neoplasms as a high risk of sternotomy procedure due to the chance of
extra-thyroidal extension of the tumour and/or the need to perform dissection of mediastinal lymph nodes [10].

The most important predictive factor as to whether a goiter can safely be removed through a cervical approach is the presence of a clear tissue plane around the nodule in the mediastinum on preoperative imaging. If such a clear plane is not present, preparations should be made for sternotomy [8]. Burns et al. performed a sternotomy in only 3/140 patients with SSG, since, in their opinion, the more significant factors giving rise to suspicion of the need to perform sternotomy are CT evidence of adherence to the surrounding mediastinal tissues and extension of the goitre to, or below, the aortic arch [22]. White et al., based on a systematic review of the literature, suggested that sternotomy is more likely to be performed in the presence of a primary substernal goiter or a mass larger than the thoracic inlet [10]. Ahmed et al. used extension beyond the aortic knuckle on chest x-ray as their landmark for the depth of substernal extension [23]. The best predictive factor for inability of the goiter to fit through the thoracic inlet was inferior extension of the goiter beyond the level of the aortic arch, as determined on either CT scan or MRI scan.

The weight of the specimen is, however, a post-operative finding and may not indicate pre-operatively the approach needed. Nevertheless, pre-operative estimation of thyroid volume, by means of CT scan, can be an effective predictor of which patients are likely to require a thoracic approach [10]. If the nodule is too large to safely traverse the thoracic inlet, sternotomy is indicated. De Perrot et al. reported that, goiters larger than 10 cm, required sternotomy. However, as they accrued additional experience, only goiters larger than 15–20 cm needed sternotomy [11].

Primary intra-thoracic or ectopic goiters present as mediastinal tumors. Ectopic goiters sometimes cannot be reached through the neck, their presence can mandate sternotomy. A minority of substernal goiters descend into the posterior mediastinum. These goiters are
difficult to extract through a cervical approach alone, and even sternotomy affords suboptimal exposure at times. Posterior mediastinal extension can, therefore, sometimes require thoracic approach in addition to a cervical incision.

The potential need for sternotomy can be predicted on the basis of preoperative CT or MRI scan. Such imaging can identify advanced malignancy with loss of the plane of dissection surrounding the goiter. Significant retrosternal extension can also be identified, along with posterior mediastinal involvement and the presence of ectopic nodule. The presence of such findings on preoperative imaging can help the surgeon prepare for the possibility of sternotomy. Median sternotomy is a standard thoracic procedure, which adds little to the morbidity of substernal goiter surgery.

Conclusions

Removal of a substernal goiter is a challenging surgical procedure. The vast majority of substernal goiters can safely be removed through the neck, but select patients are best managed via a combined cervical and sternotomy approach. The potential need for sternotomy can be predicted on the basis of preoperative CT or MRI scan. The most significant criteria for selecting patients requiring sternotomy are CT evidence of massive size of thyroid gland and extension of the goitre to or below the aortic arch and tracheae carina, in particular presence of an ectopic goitre located deep in the mediastinum, posterior mediastinal spread, malignnancy and adherence to surrounding mediastinal structures. Therefore, the CT scan should always be included in the pre-operative diagnostic workup, when substernal goiter is suspected. All these criteria can be suspected on the basis of preoperative imaging and allow the surgeon to be prepared to proceed with sternotomy, if required. However, the final decision as to whether to perform sternotomy can be made only intra-operatively, and the choice is related to the experience of the surgeon.
Conflict of interest

The authors declare that they have no sponsorship or funding arrangements relating to their research and no conflicts of interest.

References


**Figure Legends**

**Figure 1:**

A: Chest X-ray showing deviated trachea

B: Sagittal plain CT scan showing substernal goiter extension in to the anterior mediastinum

C: Coronal plain CT scan showing substernal goiter displacement mediastinal structures

D: Axial plain CT scan showing substernal goiter

**Figure 2:**

A: Coronal CT scan showing substernal goiter exerting significant tracheal deviation

B: Coronal plain MRI scan showing substernal goiter

C: MR angiography showing major vessels displacement by a substernal goiter

D: Sagittal plain MRI scan showing substernal goiter

**Figure 3:**

A: Chest X-ray showing enlarged upper mediastinum due to substernal goiter

B: Sagittal plain CT scan showing substernal goiter in the anterior mediastinum

C: Computed tomography showed that the substernal thyroid mass extends distal to the tracheal bifurcation

D: Thyroid scintigraphy showing large intrathoracic goiter