Retrosternal Thyroid Goitre Aetiology, Presentation and Management

J. Wauchope[1], T. Moran[1], E. Stanley[2], H. Rowley[1], F. O'Duffy[1,3]

1. Department of Otolaryngology and Head and Neck Surgery, Mater Misericordiae University Hospital, Dublin 7.
2. Department of Radiology, Mater Misericordiae University Hospital, Dublin 7.
3. University College Dublin, Professor of Otolaryngology Head and Neck Surgery.

Abstract

Aims
The aim of this study was to review the presentation, indications for surgery and surgical approaches to retrosternal thyroid goitres in our tertiary head and neck centre.

Methods
A retrospective 5-year review of patients who underwent surgery for retrosternal goitres (RSG) was performed from 2015 to 2020. Patients were identified through review of the HIPE database and theatre logbooks. Data was then recorded from electronic records and radiological investigations. A literature review was carried out in pubmed using the following search terms 'retrosternal goitre', 'substernal goitre', 'thyroid goitre' and 'mediastinal goitre'.

Results
32 patients were identified who received total thyroidectomy operations for RSG within this 5-year period. 27 patients (84%) had their goitres excised entirely via a transcervical approach. 1 patient (3%) required a full sternotomy and 4 patients (12.5%) required surgical intervention at the manubrium.

Discussion
The majority of RSGs can be successfully excised through a transcervical approach. A small proportion, however, may require thoracic intervention to enable removal. These intrathoracic approaches range from medial resection of clavicle, partial sternotomy to full median sternotomy. Predictive factors for thoracic intervention include posterior mediastinum involvement and extension beyond the arch of aorta. To ensure successful surgery, one needs a full armamentarium of surgical techniques.

Introduction

A goitre is defined as an enlargement of the thyroid gland to twice its normal size[1-2]. It has been estimated to affect 10% of the UK population and 1.5 billion people globally[3]. RSGs are most frequently benign with only 3-15% identified as malignant[1]. Of the benign causes iodine deficiency gives rise to the majority, however, other associated risk factors for developing the disease include increase in serum thyroid-stimulating hormone (TSH) level, natural goitrogens, smoking, and selenium and iron deficiencies[4].

With iodine deficiency being the largest causative agent of multinodular goitres, particular attention is payed to levels of dietary iodine intake globally. In 1990 the Joint UNICEF/WHO committee urged all countries to adopt and implement universal salt iodisation (USI). The introduction of USI, represented a simple intervention of an iodinated substance to table salt which did not require a change in dietary habits[5]. Most countries have taken a voluntary approach to this which has culminated in a wide variation in the production and use of iodised salt. Ireland and the UK have some of the worst figures, with iodised salt comprising only 3.3% of salt sold in Ireland and similar figures in the UK[5]. This is of clinical significance as studies have demonstrated that dietary iodine intake in Irish women remains well below WHO recommendations and Ireland is an area of borderline dietary iodine deficiency[5]. In recent years, there has been limited published data regarding the incidence of goitres within the Irish population, however with iodine deficiency still a national problem one can surmise goitres remain an endemic disease in Ireland.

Dietary goitrogens can also cause thyroid goitres, these are naturally occurring chemicals found in many plant-based foods. High consumption of these foods on a regular basis may affect your thyroid hormone production in several ways. Firstly, they can inhibit the process by which iodine is incorporated into the thyroid hormones thyroxine (T4) and triiodothyronine (T3) and secondly inhibit the release of thyroid hormones from the gland. The two main food groups that are classified as dietary goitrogens include cyogenic and flavonoid containing plant foods. These include cruciferous vegetables, starchy plants and certain fruits e.g. broccoli, kale, cauliflower, strawberries, sweet potatoes and tofu[6].

Selenium, iron and smoking have also been noted to affect the thyroid gland. Selenium deficiency decreases the synthesis of thyroid hormones which in turn increases TSH level[7]. This imbalance in thyroid hormones can result in fibrosis of the gland and goitre formation. Similarly, iron deficiency can contribute to the development of hypothyroidism and cigarette smoke contains goitrogens including thiocyanates which may lead to goitre formation[7].

Retrosternal, substernal and intrathoracic goitres are terms used to describe a subgroup of goitres that extend into the thoracic cavity[3]. Although there are several descriptions of RSG in the literature, the two most common definitions are a "thyroid gland that descends below the plane of the thoracic inlet" or a "thyroid gland with greater than 50% below the thoracic inlet"[1,2,8-10].

Retrosternal goitre (RSG) can be classified as Primary when the intrathoracic thyroid mass arises from aberrant thyroid tissue in mediastinum and is entirely separate from the neck and Secondary RSG which result from the downward growth of a normally situated thyroid gland in the neck[1].

Goitres may present to surgeons for diagnostic and therapeutic purposes. Of all multinodular goitre patients undergoing total thyroidectomy, the incidence of RSGs has been reported to be between 1-20%. The wide variation in reported incidence has been partially surmised to be due to the lack of consensus regarding the definition of RSG[11]. The diagnosis is usually made in the 5th-6th decades of life with a female preponderance 4:1[11]. The majority of RSGs (85-90%) are located within the anterior mediastinum, with the remainder (10-15%) located in the posterior mediastinum[11].

They can cause a variety of symptoms including respiratory distress, dysphagia and thoracic inlet obstruction. These symptoms are induced by extrinsic compression of the trachea, oesophagus and great vessels as the goitre grows in size[1]. The gold standard treatment for symptomatic retrosternal goitres is thyroidectomy. There is no substantial supporting evidence in the literature for the use of medical suppression therapy in the form of levothyroxine or radioactive iodine[1].

We aimed to review the presentation, indications for surgical intervention and surgical techniques in retrosternal thyroid surgery within our own institution.

Methods

A literature review was carried out in pubmed using the following search terms 'retrosternal goitre', 'substernal goitre', 'thyroid goitre' and 'mediastinal goitre'. Following this, a retrospective 5-year review of patients who underwent surgery for retrosternal goitres was performed in our tertiary hospital. After discussion with our institution's ethics committee, ethical approval was deemed not to be required as this review was primarily an audit. All patients who received total thyroidectomy operations were identified with the assistance of the Hospital In-Patient Enquiry (HIPE) Department and by reviewing theatre logbooks from 1st of January 2015 to 1st of January 2020. A small proportion of thyroidectomy surgery was still undertaken by General Surgeons previously, however, in recent years this operation has been solely undertaken by ENT in our institution. The hospital's electronic patient care record was then reviewed for each patient looking at relevant outpatient clinic letters. Patients with symptomatic RSGs were investigated with CT neck/thorax imaging to assess the size, location and compressive effects of the goitre. For the purposes of this study a retrosternal goitre was defined as a thyroid goitre confirmed radiologically to be extending beyond the thoracic inlet.

Preoperative CT imaging identified maximal retrosternal area of each goitre and its relationship to the trachea, oesophagus and major vessels. In patients with asymptomatic or mildly symptomatic retrosternal goitres we routinely elect for a period of observation with a repeat CT neck/thorax on presentation or progression of symptoms.

The presence of positive symptoms and/or time to progression of symptoms were documented for each case. Following this, operative notes were examined to identify surgical approach and technique for each patient's procedure.

Results

A total of 32 patients were identified who received total thyroidectomy operations for retrosternal goitres within this 5-year period under ENT in our institution, which consisted of four separate consultants. The indication for surgery was compressive symptoms secondary to the goitre in all 32 patients. There was a female preponderance of 3.5:1, with 25 of the 32 patients being female. The age of patients undergoing surgery ranged from 30 to 85 with a median age of 65. The most common symptoms reported were orthopnoea, dyspnoea, choking/pressure sensation and dysphagia respectively (see figure 1). Seventy eight percent of patients reported varying degrees of orthopnoea, whilst only 9% had stridor and 6% were documented to be Pemberton's positive on clinical examination.

Figure 1: Table showing occurrence rates of clinical symptoms/signs.

| CLINICAL SYMPTOMS/SIGNS	PERCENTAGE (Number)
Orthopnoea	78% (25)
Dyspnoea	50% (16)
Choking/pressure	47% (15)
Dysphagia	47% (15)
Stridor	9% (3)
Hoarseness	6% (2)
Pemberton's positive	6% (2)
Cough	3% (1)

The radiological retrosternal extent of goitre was recorded in 23 patients. With a retrosternal extension range beyond the manubrium of 2cm – 14 cm with an average of 7.5cm. All of these patients had radiological evidence of airway compromise with tracheal deviation and/or compression. The degree of tracheal compression was reported in 18 patients. The range of reduction of tracheal lumen was 3mm – 15mm with an average reported compression of trachea to 7.8mm (see figure 2).

Figure 2: CT thorax in coronal plane depicting a retrosternal goitre causing significant tracheal compression.

Length of time from onset of symptoms to initial review and surgery date was ascertained. Twenty eight out of the thirty two patients had retrievable figures on this data point. Of these 28 patients, length of time from onset of symptoms to surgery ranged from 0-144 months, with an average of 34 months (see figure 3). Length of time from initial ENT review to surgery ranged from 0-132 months with an average observation time of 11.7 months. One patient had immediate surgery during acute presentation to the emergency department with stridor.

Figure 3: Graph showing length of time in months from onset of symptoms to surgery date (x axis shows case number and y axis is time in months).
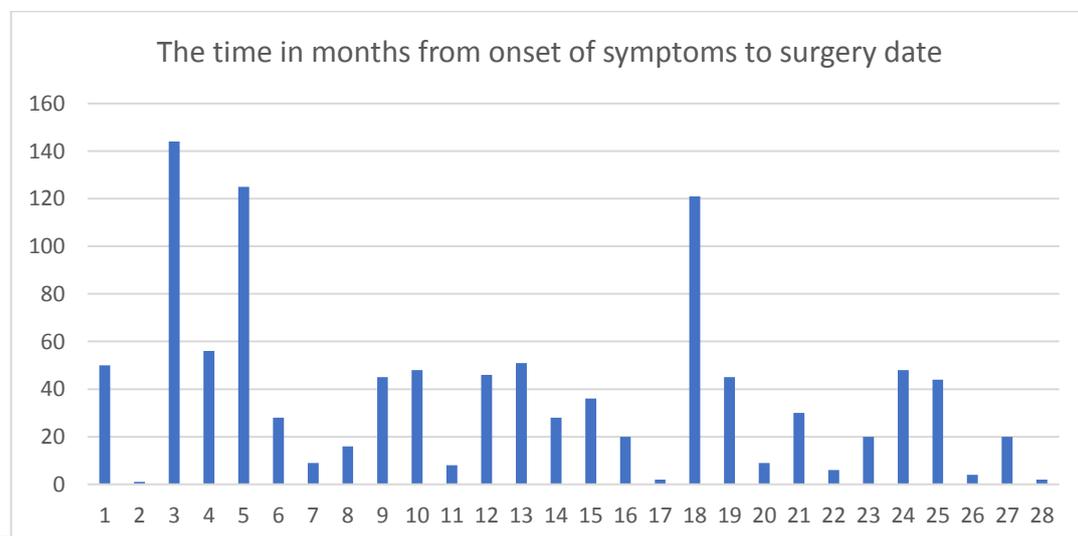
Once decision has been made to proceed with thyroidectomy, a Cardiothoracic Surgery consult is requested for those patients deemed high risk of requiring a sternotomy. This is decided on an individual patient by patient basis by reviewing each patient's clinical condition and pre-operative cross-sectional imaging and noting the presence of any positive predictive factors for requiring sternotomy. For those patients deemed high risk of requiring sternotomy, Cardiothoracics team remain on standby for the surgery.

For the 32 patients identified, 27 patients (84%) had their goitres excised entirely via a transcervical approach. 1 patient (3%) required a full sternotomy and the remaining 4 patients (12.5%) required surgical intervention or manoeuvres at the thoracic inlet.

Surgical Technique

A skin crease incision is made 2-4 cm above the sternal notch. Subplatysmal flaps are then raised and the sternothyroid muscle is routinely divided. The cervical thyroid mobilised and thyroid vessels are ligated. Once the cervical thyroid is mobilised, we assess the mobility of retrosternal component and decide on need for additional access at the thoracic inlet. Thoracic inlet techniques described in these five cases included soft tissue split down to the sternal notch with vertical midline incision and (see figure 4), division of the sternohyoid muscle, manubriumectomy to increase diameter of thoracic inlet, partial upper sternotomy or total sternotomy. For the patients who required partial upper and total sternotomies, it was performed by Cardiothoracics Surgery team.

Figure 4: Intraoperative image of a retrosternal goitre after being delivered through a transcervical approach with an extended midline incision and soft tissue split down to the sternal notch.

Post operatively all patients were extubated and with no symptoms of tracheomalacia and none required a tracheostomy. Two patients (6%) had prolonged complications of hypocalaemia, however, none of the 32 patients (0%) suffered from permanent recurrent laryngeal nerve injury.

Discussion

Goitres can cause significant compressive symptoms including respiratory distress, dysphagia and vascular compromise of the major vessels[11]. The literature reports that retrosternal goitres are more likely to result in varying degrees of pressures effects, compared with purely cervical goitres, which may culminate in acute respiratory compromise and sudden death[12]. Tracheomalacia has been reported in 1% and superior vena cava syndrome in 3.2% of patients with RSGs according to one systematic review[8]. As there is no reliable medical management, surgery remains the treatment of choice for retrosternal goitres with or without clinical symptoms due to the apparent life-threatening risks of an enlarging goitre[1,2].

The slow growing nature of thyroid goitres is an important consideration in patient selection for surgery. The risk-benefit profile for elderly patients with co-morbidities i.e. life expectancy measures in years rather than decades, often does not favour surgical intervention but rather observation to ensure no emergent intervention is required for compressive symptoms. The average time from presentation to surgery of 11.7 months reflects the authors considered approach to intervention, with a period of observation of symptoms and serial imaging to assess need for surgical intervention.

The vast majority of RSGs can be successfully excised via a cervical approach with thoracic intervention required in only a small subgroup reported between 0-11%[11], with one paper stating that all RSGs could be removed via the neck[13]. A small proportion of these patients, however, may require additional intervention, which is not standard practice for thyroidectomy operations, to enable removal. These techniques range from less extensive procedures such as medial resection of the clavicle and manubriumectomy; to more invasive approaches including full median sternotomy and lateral thoracotomy. The literature has reported a significant association between the extent of RSG and reported complications, with the incidence of tracheomalacia, superior vena cava syndrome and the need for thoracic approach increasing 10-fold in patients with RSGs extending to the aortic arch[8]. The predictive factors reported for extra-cervical approaches include involvement of the posterior mediastinum, extension beyond the aortic arch, previous goitre surgery, superior vena cava obstruction, malignancy and emergent airway obstruction[8,11,14-16].

The greatest operative risk when resecting an intrathoracic goitre is the potential for catastrophic thoracic haemorrhage which the surgeon would not be able to control due to lack of access to thoracic vessels. The potential for sternotomy or manubrium manoeuvres can and should be anticipated depending on the features of the goitre as previously described. In order to optimize patient safety and avoid the scenario of an uncontrolled haemorrhage it is paramount that these high-risk goitres be operated in a surgical centre with the ability to undertake a sternotomy.

Patients may also require a tracheostomy post operatively due to bilateral vocal cord palsy or significant tracheomalacia. To ensure successful retrosternal thyroid surgery, one needs a full armamentarium of surgical techniques, equipment and specialists.

Declaration of Conflict of Interest:
The authors have no conflicts of interest to disclose.

Corresponding Author:
Jessica Wauchope
Mater Misericordiae University Hospital,
Dublin 7.
E-Mail: jwauchope02@qub.ac.uk

References:
1. Hashmi S, Premachandra D, Bennett A, Parry W. Management of retrosternal goitres: results of early surgical intervention to prevent airway morbidity, and a review of the English literature. The Journal of laryngology and otology. 2006;120(8):644.
2. Sheng YR, Xi RC. Surgical approach and technique in retrosternal goiter: Case report and review of the literature. Annals of medicine and surgery. 2016;5:90-2.
3. Simó R, Nixon IJ, Vander Poorten V, Quer M, Shaha AR, Sanabria A, et al. Surgical management of intrathoracic goitres. European Archives of Oto-Rhino-Laryngology. 2019;276(2):305-14.
4. Knobel M. Etiopathology, clinical features, and treatment of diffuse and multinodular nontoxic goiters. Journal of Endocrinological Investigation. 2016;39(4):357-73.
5. Nawoor Z, Burns R, Smith D, Sheehan S, O'herlihy C, Smyth P. Iodine intake in pregnancy in Ireland—a cause for concern? Irish journal of medical science. 2006;175(2):21.
6. Chandra AK. Chapter 42 - Goitrogen in Food: Cyanogenic and Flavonoids Containing Plant Foods in the Development of Goiter. Elsevier Inc; 2010. p. 691-716.
7. Hughes K, Eastman C. Goitre: Causes, investigation and management. Australian family physician. 2012;41(8):572.
8. Huins CT, Georgalas C, Mehrzad H, Tolley NS. A new classification system for retrosternal goitre based on a systematic review of its complications and management. International Journal of Surgery. 2008;6(1):71-6.
9. DeSouza F, Smith P. Retrosternal goiter. The Journal of otolaryngology. 1983;12(6):393.
10. Nankee L, Chen H, Schneider DF, Sippel RS, Elfenbein DM. Substernal goiter: when is a sternotomy required? Journal of surgical research. 2015;199(1):121-5.
11. Coskun A, Yildirim M, Erkan N. Substernal goiter: when is a sternotomy required? International surgery. 2014;99(4):419-25.
12. Singh B, Lucente FE, Shaha AR. Substernal goiter: a clinical review. American journal of otolaryngology. 1994;15(6):409-16.

13. Netterville JL, Coleman SC, Smith JC, Smith MM, Day TA, Burkey BB. Management of substernal goiter. The Laryngoscope. 1998;108(11):1611-7.

14. Kang C, Williams L, Arora A, Dae K, Enyinnaya O. Retrosternal goitres–Predictors for sternotomy. European Journal of Surgical Oncology. 2017;43(12):2386.

15. Casella C, Pata G, Cappelli C, Salerni B. Preoperative predictors of sternotomy need in mediastinal goiter management. Head & neck. 2010;32(9):1131-5.

16. Hanson MA, Shaha AR, Wu JX. Surgical approach to the substernal goiter. Best Practice & Research Clinical Endocrinology & Metabolism. 2019;33(4):101312.