A new classification system for retrosternal goitre based on a systematic review of its complications and management


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Abstract

Introduction: There is no standard definition for thyroid glands extending below the thoracic inlet, and there are no clear guidelines for pre-operatively identifying those patients that may require an intrathoracic approach.

We therefore reviewed the current literature in order to establish the current practices regarding the management of retrosternal goitres (RSGs), and propose a classification system to aid pre-operative planning for this important group of patients.

Materials and methods: A PubMed Medline search was conducted using the search terms 'retrosternal', 'substernal', 'intrathoracic', 'mediastinal', 'goitre' and 'goiter', resulting in 626 hits. Exclusion criteria reduced the number of papers to the 34 used for this review.

Results: A total of 34 papers totaling 2426 patients were included. Eighty-four percent of patients operated on for RSG were achieved via a cervical approach, with the remainder also requiring manubriotomy (3.1%), full sternotomy (6.6%) or thoracotomy (4%). Tracheomalacia occurred in 1% of patients and Superior Vena Cava syndrome (SVC) in 3.2%. There was a clear and highly significant association between the extent and definition of RSG and reported complications, as well as the approach used, with the incidence of tracheomalacia, SVC and need for intrathoracic approach increasing more than 10-fold in cases of RSG reaching the aortic arch.

Discussion: There is a clear need to establish a common standard in the definition and description of the extent of RSG. Using our findings, we propose a new, simple, 3-grade classification system of RSGs, based on their relation with the aortic arch.

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An enlarged thyroid can be the clinical manifestation of several pathologies, although most commonly it is associated with dietary iodine deficiency leading to subclinical hypothyroidism, with subsequent elevated TSH and gland hypertrophy.

Terms such as retrosternal, substernal, intrathoracic, or mediastinal have been used to describe a goitre that extends beyond the thoracic inlet. However, there is lack of consensus regarding the exact definition of a retrosternal goitre (RSG). Controversies exist regarding the incidence of tracheomalacia in RSG, with its anaesthetic and surgical implications. Superior vena cava syndrome has been associated with RSG, although its incidence and significance vary widely between studies. Even more importantly, from a surgical point of view, the approach required for an RSG is the subject of significant debate. Although various classification systems for RSG have been proposed, none of these offer a correlation between size and/or location of the RSG and the extent of surgery that may be necessary.

We feel that the significant variability found in the literature regarding the clinical presentation and the surgical management of RSG can — at least in part — be explained by the different definitions employed. With this in mind, we performed a structured review of the complications and management of RSG, accounting for the definitions used by different authors and, on the basis of our findings, suggest a new classification system designed to improve communication of findings and results between clinicians as well as assist in preoperative planning.

Materials and methods

The Cochrane Controlled Trials Register (CCTR), Medline and EMBASE were searched from 1966 onwards. The search was performed in March 2006 using different combinations of the expanded search terms ‘retrosternal’, ‘substernal’, ‘intrathoracic’, ‘mediastinal’, ‘goitre’ and ‘goiter’ and 626 articles were found. No controlled trials were found. After excluding case reports, basic science articles and correspondence, 34 papers were used for the purpose of this review, 29 of which were case series. Data extracted from the series was transferred to SPSS (13.0) and analysed. Chi-square test and Fisher exact tests were used for all comparisons in proportions.

Results

On review of the abstracts available, 34 recent papers were selected for their relevance and their full texts obtained. Five papers were reviews, which were excluded from the calculations; 29 included papers were case series. The number of patients in each study ranged from 18 to 7480, with a total of 2426 patients identified as having a RSG.

Definition

Four definitions of RSG were identified: (1) a thyroid in which any part of the gland extends below the thoracic inlet with the patient in the surgical position; (2) a gland reaching the level of the aortic arch; (3) a thyroid reaching the level of T4 (on chest x-ray) and (4) greater than 50% (or ‘the majority’) of the gland residing below the thoracic inlet, first proposed by deSouza and Smith (Table 1).

<table>
<thead>
<tr>
<th>Below thoracic inlet</th>
<th>Aortic arch</th>
<th>T4</th>
<th>&gt;50% below thoracic inlet</th>
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<tr>
<td>Kaya et al. (1994)</td>
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<td>Bennett et al. (2004)</td>
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<td>Flati et al. (2005)</td>
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Total papers: 8 Total paper: 1 Total paper: 1 Total papers: 23
Total patients: 1031 Total patients: 175 Total patients: 24 Total patients: 1109
(24 patients); in one\textsuperscript{21}, no definition was specified. It is interesting to note that although less than one fourth of all papers used the rather loose definition of “any part of the gland below the thoracic inlet”, these papers accounted for almost half (44%) of all patients.

**Incidence**

Using the data from those 17 papers that stated an incidence of RSG, the cumulative incidence of retrosternal extension was 1389 (6.28%) out of a total of 22,125 patients with a goitre. Previous surgery had taken place in 1.7\textsuperscript{e}30\% of cases, with sub-total thyroidectomy being the most common first procedure.

**Approach**

Out of a total of the 2426 patients operated on for RSG, in 1745 (71.9\%) the cervical approach was sufficient. Sixty-five patients (2.7\%) required additional manubriotomy, 131 (5.4\%) full sternotomy and 91 (3.6\%) required a form of thoracotomy such as Hart’s procedure (Table 2).

The highest incidence of manubriotomy was found in those RSGs defined as reaching the aortic arch (27 out of 175 patients, 15.4\%), followed by patients with a RSG descending to the level of T4 (1 out of 24, 4.2\%); 15 out of 1109 patients (1.3\%) in whom >50\% of the thyroid resided within the thorax required a manubriotomy. However, only 10 out of 1031 of the patients (0.97\%) with “any part of the gland below the thoracic inlet” required manubriotomy.

Eighty out of 1109 patients (7.2\%) with the majority of the thyroid below the thoracic inlet required sternotomy, followed by those whose gland reached the aortic arch (11 out of 175, 6.29\%), while only 3.7\% of the patients with RSG defined as “any part of the thyroid gland extending below the thoracic inlet” required sternotomy. Other procedures (such as a lateral thoracotomy or Hart’s procedure) were performed in 91 patients (3.8\%), mostly for RSGs reaching the aortic arch (41 out of 175, 23.4\%), followed by those below the thoracic inlet (27 out of 1031, 2.62\%) and finally those RSGs whose majority was below the thoracic inlet (23 out of 1109, 2.07\%). In summary, a thoracic approach was required in 79 out of 175 patients (45.1\%) with thyroid gland reaching the aortic arch, 118 out of 1109 patients (10.6\%) with more than 50\% of the gland below the thoracic inlet, and in 75 out of 1031 patients (7.2\%) in which RSG was defined as “any part of the gland below the aortic arch”. The difference was statistically significant (\textit{P} < 0.001) (Table 2).

**Complications**

Specific complication rates were quoted with regard to RSG in 107 out of 2426 patients (4.41\%), including Superior Vena Cava syndrome in 67 patients (2.7\%) and tracheomalacia in 21 patients (0.8\%). Tracheostomy was performed in 19 patients (0.78\%) (Table 3).
Combining these results with the approach used, the highest incidence of tracheostomies was seen in patients with RSG reaching the level of the aortic arch (4 out of 175, 2.29%), followed by 13 performed for those with the "majority of the RSG within the thoracic cavity" (13 out of 1109, 1.17%) but only 2 out of 1031 (0.19%) "below the thoracic inlet". Similarly, tracheomalacia occurred only in those RSGs descending to the aortic arch (18 out of 175, 10.3%) and those in which the majority of the RSG resided within the chest (3 out of 1109, 0.27%); none occurred in RSGs below the thoracic inlet, or those reaching T4. Again, Superior Vena Cava syndrome was commonest in those RSGs reaching the aortic arch (21 out of 175, 12%) followed by those whose majority resided within the chest (28 out of 1109, 2.52%), and then those below the inlet (18 out of 1109, 1.62%); it was not described in those descending to T4. The correlation between tracheomalacia, SVC syndrome and the extent of RSG was statistically highly significant (P < 0.001).

Of the 19 patients requiring tracheostomy, it was performed in six (32%) for persistent tracheomalacia, in three (16%) for pre-existing bilateral vocal cord palsy, in two (11%) as a result of unresectable malignancy, in one patient it was performed for a tracheal–laryngeal stricture and one following a post-operative haematoma; three (16%) were stated as "not complications per se", and reasons for a further three (16%) were not specified.

Tracheomalacia is defined as "softening of the tracheal cartilages ... usually accompanied by a barking cough and expiratory stridor or wheezing". Traditionally, this has been treated with conservative methods such as prolonged intubation or positive pressure ventilation, or more invasive methods including placing of support sutures, use of a tracheal stent, or even tracheostomy. In this review, when this was encountered, it was treated with prolonged intubation in 14 patients (67%), sutures were placed in two patients (10%), one (5%) received a tracheal stent and six patients (29%) underwent a tracheostomy.

Discussion

There is much controversy concerning the definition of retrosternal goitres. Cougard et al. regarded any part of a goitre extending below the thoracic inlet as retrosternal. Cire in 1939, and others since, offered definitions of varying depths into the thoracic cavity, whilst the most common definition, proposed by deSouza and Smith, defined a retrosternal goitre as that in which >50% of the total bulk of thyroid tissue resides below the thoracic inlet. All of these definitions, however, can be interpreted rather loosely, and lack the anatomic clarity gained by referring to an intrathoracic landmark, such as the aortic arch. Although there are arguments for and against the use of each definition, it is clear that until we develop a common language, we will be unable to compare our patients and results: patients with a thyroid gland barely below the thoracic inlet are a very different group of patients from those with a thyroid gland reaching the aortic arch. The fact that this is more than just of academic interest is highlighted in our results.

Although tracheostomy was performed in only 0.8% of all the patients with RSGs, there was a clear and highly significant trend of increasing incidence moving from "any part of the gland below the inlet" to "the majority of gland below the thoracic inlet" to "at the level of the aortic arch". Exactly the same pattern was evident in the case of tracheomalacia, where the incidence rose from 0% to 10%, in cases of RSGs reaching the aortic arch. However, only one patient had a stent inserted, suggesting that significant tracheomalacia, whilst not a myth as claimed by Bennett et al., is extremely rare; indeed, this is certainly the experience of the senior author. SVC syndrome is a rare complication, and again, its incidence closely mirrors that of tracheomalacia — virtually nonexistent in thyroid glands extending just below the thoracic inlet, but rising to more than 10% in RSGs extending to the level of the aortic arch.

The cervical approach can be utilized in the majority of cases, with one paper claiming that all RSGs can be excised via this route. However, cervicotomy can increase the inherent risks of surgery due to poor access. For cases in which the cervical approach is not adequate, ‘intrathoracic’ approaches have been employed, including manubriotomy, full sternotomy and lateral thacotomy. Manubriotomy, or full sternotomy, have many advantages over lateral thacotomy. They have been proven in cardiac surgery to employ an incision that can be performed quickly, reliably and with very low morbidity, providing excellent mediastinal exposure to allow delivery of the retrosternal portion of the gland. Moreover, it can be performed without having to reposition the patient. On average, median sternotomy increases the hospital stay by only two days over cervical thyroidectomy.

Sand et al. have proposed indications for sternotomy including pressure effects, recurrent retrosternal goitres, potential for acute airway problems, malignant potential, severe venous obstruction, primary (isolated retrosternal) goitres, retrosternal goitres reaching the level of the aortic arch, anticipation of the need for excessive traction to expose the recurrent laryngeal nerves, or if the lowermost extent of the tumour cannot be palpated.

CT scanning has become the gold-standard pre-operative radiological investigation. Grainger et al. reviewed their practice to identify whether a certain group of patients were more likely to require a median sternotomy based on CT findings. Their retrospective study looked at 27 patients who underwent surgery for RSG, 24 of whom had pre-operative CT scans. Three patients (12%) required a median sternotomy in addition to mobilize the gland. They have been proven in cardiac surgery to employ an incision that can be performed quickly, reliably and with very low morbidity, providing excellent mediastinal exposure to allow delivery of the retrosternal portion of the gland. Moreover, it can be performed without having to reposition the patient. On average, median sternotomy increases the hospital stay by only two days over cervical thyroidectomy.

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On the basis of our findings, we suggest a new classification system for RSG; as has been previously stated, an indication for an intrathoracic approach is the inability to reach the inferior aspect of the gland via a cervical incision alone. Therefore, and as reflected in our studies, it would seem inappropriate to perform a full sternotomy for...
retrosternal goitres that do not reach to the level of the aortic arch. However, extension to and below the aortic arch and to the level of the pericardium cannot be reached safely via a cervical approach alone, requiring some degree of intrathoracic approach to gain adequate exposure and allow delivery of the intact gland, which can be attained by manubriotomy as shown by Monchik and Marerazzi. Any gland below the level of the right atrium would require a full sternotomy for adequate exposure. These levels can therefore be classified as levels 1, 2 and 3, respectively (see Table 4). Given that retrosternal goitres are essentially a midline structure, together with the documented ease of performance, excellent exposure and minimal associated morbidity, sternotomy would seem the obvious approach rather than a lateral thoracotomy. This classification system correlates with the results from Grainger et al. with regard to findings on CT scanning and, more importantly, reflects to a significant degree the practice of the majority of surgeons, as shown in our study.

In highlighting these findings on CT scanning, the appropriate preparations can be made for grade 2 or 3 RSGs, be it referral to an appropriate hospital with intrathoracic capabilities, or liaison with cardiothoracic colleagues to employ a multidisciplinary team approach.

In summary, we feel that this classification system, used together with a uniform definition for retrosternal goitres, will provide a common standard for pre-operative planning of the management of these patients, and one against which results can be compared. By planning, we can avoid any unexpected requirement for intrathoracic extension intra-operatively, the unnecessary use of resources in cases planned for an intrathoracic approach when one was not required, and morbidity associated with inappropriately performed procedures. Finally, the appropriate consent of the patient can be taken in cases where an intrathoracic approach may need to be employed, so that the patient can make a fully informed decision regarding the proposed surgery.

Conflicts of interest
None.

Funding
None.

Ethical approval
Not required.

References

Table 4 Classification and approach for retrosternal goitres

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<thead>
<tr>
<th>Grade</th>
<th>Anatomical location</th>
<th>Approach</th>
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<tr>
<td>1</td>
<td>Above aortic arch (above T4)</td>
<td>Cervical</td>
</tr>
<tr>
<td>2</td>
<td>Aortic arch to pericardium</td>
<td>Manubriotomy</td>
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<tr>
<td>3</td>
<td>Below right atrium</td>
<td>Full sternotomy</td>
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