A goitre is defined as an enlargement of the thyroid gland to twice its normal size or to a weight of more than 40 grams (1). The first substernal goitre was described by Haller in 1749 (2). In 1820, Klein performed the first ever surgical resection of a substernal goitre (3). Since then, numerous studies have been published and a myriad of definitions for substernal or intrathoracic goitres have been proposed, according to individual surgeon’s clinical and radiologic criteria. Ríos et al. analysed ten such definitions, concluding that all but two were not clinically relevant (4). The first is the clinical definition, which describes “a thyroid gland that, on neck examination without being in hyperextension, has a portion that remains permanently retrosternal”. The second is Katlic’s definition which describes a “goitre in which at least 50% is retrosternal”, thus useful in predicting the need for a sternotomy (5).

Mediastinal goitres are classified as primary or secondary. Primary mediastinal goitres are those without any direct fibrous or parenchymal connection to the cervical portion of the gland. Further definitional criteria include blood supply from a mediastinal source, a normal or absent cervical thyroid gland, no history of prior thyroid surgery and a lack of similar pathology in the other portions of the thyroid gland. Primary mediastinal goitres represent less than 1% of intrathoracic goitres (1). Secondary goitres are far more common, defined as those developed by the downward growth of the cervical gland, with a blood supply derived from cervical branches of the thyroid arteries. The incidence of secondary mediastinal goitres varies substantially in the literature from 2% to 20%, due to the variation in used definitions. Most secondary goitres grow into the anterior mediastinum, whereas only 10–15% lie in the posterior mediastinum (1,5).

The majority of mediastinal goitres are diagnosed in the sixth decade of life with a female to male ratio of 3:1. Amongst them, 20% to 30% are barely palpable in the neck, whereas approximately 40% are diagnosed incidentally. If symptoms are present, these are related to compression of the airways or oesophagus. Dyspnoea, sleep disturbance,
dysphagia and hoarseness are the most common symptoms described in the literature. Most patients tend to have normal thyroid function, although there have been cases of hyper- or hypothyroidism, which can be detected on the basis of laboratory testing, including blood tests (1,5). Recently, a retrospective study examining 140 patients with retrosternal goitre revealed 112 (80%) euthyroid patients, while only 28 (20%) exhibited hyperthyroidism (6). Similarly, about the same percentage of hyperthyroidism (18.5%) was noticed in another cohort of patients diagnosed with intrathoracic goitre, while the rest had normal thyroid function (7). Computed tomography (CT) scanning of the neck and chest is the standard tool of investigation to ascertain diagnosis. Even in asymptomatic cases, it is generally agreed that surgical treatment is required given the potential airway compromise and ineffectiveness of non-operative treatment. In their systematic review, White et al. revealed that the incidence of malignancy in substernal goitres is similar to the one found in cervical goitres (3% to 21%) (8). Previous radiotherapy, presence of cervical adenopathy, recurrent goitre and family history of thyroid pathology have been described as risk factors for malignancy.

Although surgical management is agreed upon, the indications for an extracervical approach still remain controversial. According to White et al., the main problem lies in the different criteria used to define a substernal goitre in the literature. Therefore, most of the data comes from a heterogenic series, rendering them incomparable (8). In general, a cervical approach is sufficient for removing the vast majority of substernal goitres. Among experienced endocrine surgeons, the percentage of an extracervical approach lies at the level of 2% (8); however, many surgeons have reported rates of 11% (9) or eventually 31% in the past (10). A transcervical approach to mediastinal tumours, in particular those located in the posterior mediastinum, can be difficult. Ehrenhaft and Buckwalter reported a higher risk of uncontrollable haemorrhage, injury to the recurrent laryngeal nerve (RLN) and incomplete removal of the goitre with transcervical approach used for resection of thoracic goitres (11). Generally, a cervical approach combined with sternotomy is favoured for resection of anterior mediastinal goitres, while thoracotomy is advised for removal of posterior mediastinal goitres. Van Schil et al. highlighted thoracotomy for the removal of substernal goitres as a method to avoid troublesome bleeding (12). It is noteworthy that manoeuvres involving blind dislocation of the gland from the mediastinum towards the neck (Foley catheter applied through the cervical incision, morcellation, use of heavy silk structures into the cervical component to apply traction) are discouraged due to high risk of haemorrhage or damage of adjacent structures located in the thoracic inlet.

Factors reported to increase the likelihood of using an extracervical approach include: presence of a mass that is larger than the thoracic inlet or a mass inaccessible from the neck, involvement of the posterior mediastinum, extension of the goitre to the aortic arch, large thyroid tissue extending towards the tracheal bifurcation, presence of a recurrent postoperative goitre, superior vena cava compromise, preoperative diagnosis of malignancy with suspicion of involvement of neighbouring structures, ectopic thyroid tissue in the mediastinum, airway obstruction or inability to palpate the lowermost extent of the gland (8,13). The presence of a clear tissue plane is the most important predicting factor as to whether a goitre can safely be removed solely with a cervical incision (14). Although all the preoperative planning of an extracervical approach would be required, the decision to proceed with it should be made intraoperatively, after assessing the feasibility of removing the goitre using an accurate cervical approach.

In order to avoid the more aggressive approaches of thoracotomy or sternotomy in the surgical management of intrathoracic goitres, different techniques have been proposed, such as transclavicular access as described by D’Alia et al. (15). Fortunately, the use of minimally invasive techniques, such as VATS (video-assisted thoracoscopic surgery) and the da Vinci robotic surgical system have advanced tremendously in the last decade. Minimally invasive approaches are associated with faster recovery, reduced morbidity and pain, shorter hospital stay and better cosmetic results compared to open surgery.

There has been accumulating evidence that VATS may provide a reliable alternative to thoracotomy, thus exploiting the advantages of minimal invasive approaches. Shigemura et al. employed VATS along with a supraclavicular window in five, high-risk patients with huge anterior mediastinal goitres resulting in uncomplicated postoperative course and favorable outcomes in all cases (16). Gupta et al. described the use of VATS in seven cases of retrosternal goitres highlighting its potential benefits over sternotomy and thoracotomy. Additionally, Bhargav et al. recently released a series of posterior mediastinal goitres (11 cases) that were treated through thoracoscopic approach. No major morbidity was
revealed except for one case of RLN injury (17). Despite encouraging preliminary results, there are still some limitations with thoracoscopy, which may discourage some surgeons to proceed with this approach. These include the 2D visualisation provided by the system and the difficult access to the upper mediastinum due to the rigidity and length of the VATS instruments.

Robotic access may overcome some technical aspects of the VATS approach. According to Podgaetz, the system offers superior manoeuvrability and 3D visualisation, which permits a precise dissection of the delicate vessels surrounding the thyroid gland and its mediastinal extension (18). Rea et al. described their experience of 108 robotic-assisted thoracoscopic operations for mediastinal diseases, including one ectopic goitre, with no reported surgical mortality. As noted, the da Vinci system allows resection of almost all the mediastinal diseases, irrespective of their location, providing access even in remote areas (19). Furthermore, Wang et al. described the use of robot-assisted approach in the treatment of a huge retrosternal goitre (20). It has to be noted that the aforementioned studies reported on a combined cervico-mediastinal approach for posterior masses; mediastinal approach was crucial for the dissection and mobilization, whereas neck incision aided in the removal of the goitres (18,20). Even though some promising results have come to light so far, the role of the da Vinci system in thoracic surgery remains to be elucidated, given the recent advent of this technique. Certainly, two main drawbacks pose dilemmas for its establishment in clinical practice; high cost and the slow learning curve.

Complications following surgical excision of goitre vary among different institutions; however, permanent hypoparathyroidism, permanent nerve injury and tracheomalacia are some often encountered problems that need to be addressed carefully. It has been shown that removal of substernal goitres is associated with increased risk of permanent hypoparathyroidism and laryngeal nerve injury that in some cases may reach up to 5% and 14%, respectively (8,21,22). In contrast, the aforementioned complications occur only in 1% to 2% of patients undergoing total thyroidectomy for cervical goitres (8). Similarly, tracheomalacia seems to present with low incidence (0–2%) after resection of substernal goitres. Long-standing presence of the goitre, especially more than five years, and significant tracheal deviation or compression have been recognized as risk factors for tracheomalacia and tracheostomy (23). There is no consensus on the optimal treatment of tracheomalacia; however, conservative approach is usually associated with a favorable outcome (8,24).

Generally, postoperative evaluation should include close monitoring of respiration and circulation as well as recording of potential voice disturbances and symptoms related to hypocalcaemia. Serum calcium and parathyroid hormone concentrations must be evaluated on a daily basis and if dysregulation is evident, supplementation of oral calcium or calcitriol (1,25-dihydroxycholecalciferol) should be initiated. In case of voice disorder or easy aspiration on swallowing, patients should be referred to an otolaryngologist for laryngoscopic evaluation of vocal cord palsy.

In conclusion, an extracervical approach may be employed for a mediastinal goitre with more than 50% of its mass located in the mediastinum. A thoracic surgeon needs to be present in case a sternotomy (for glands located in the anterior mediastinum) or thoracotomy (for those located in the posterior mediastinum) is required. New, alternative approaches are the minimally invasive techniques, which include VATS and robotic surgery. These seem to be associated with favourable outcomes as they reduce morbidity and provide a preferable alternative to the aggressive and risky manoeuvres often used by experienced surgeons in order to avoid an extracervical approach. While thoracic surgeons are becoming more experienced in the use of VATS, this remains a demanding approach given its limitations in visualisation and instrumentation. The da Vinci surgical system offers a very attractive alternative to open surgery, as it allows precise dissection and better visualisation. Although preliminary results quote its efficacy in clinical practice, more studies need to be conducted in order to definitely establish its role in this field. Hopefully, in the future, the financial costs of robotic surgery will decrease, allowing it to become the gold standard in the management of mediastinal goitres.

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Footnote

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References