

Recent Trends in Management of Retrosternal Goiter

An Essay

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بِسْمِ اللَّهِ الرَّحْمَنِ الرَّحِيمِ

قالوا

سببناك لا تعلم لنا
إلا ما علمتنا إنك أنت
العليم العظيم

صدق الله العظيم

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List of Abbreviations

RSG	:	Retrosternal Goiter.
SEMS	:	Self-expandable metallic stent.
MAO	:	Malignant airway obstruction.
CT scan	:	Computerized tomography scan.
PET scan	:	Positron emission Tomography scan.
MRI	:	Magnetic resonant Imaging.
VATS	:	Video Assisted thoracoscopic Surgery.
RLN	:	Recurrent laryngeal nerve.
IONM	:	Intra-operative nerve monitoring.
TRH	:	Thyrotropin-releasing hormone.
T3	:	Triiodothyronine.
T4	:	Thyroxine hormone.
SVC	:	Superior vena cava.
JVP	:	Jugular venous pressure.

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Introduction

Retrosternal goiter was first described by Albrecht von Haller in 1749, as the extension of the thyroid tissue below the upper opening of the chest. Since then retrosternal goiter has always been considered a challenge for the surgeons, because of the difficulties that may be encountered during surgical removal. The definition of retrosternal goiter is still not uniform, and varies between the different Authors. However, the most commonly accepted definition of retrosternal goiter describes a goiter as retrosternal when a $\geq 50\%$ portion of the mass is located in the mediastinum (*Netterville et al., 1998*).

The presence of retrosternal goiter is documented in 2-19% of all thyroidectomies, Diagnosis of retrosternal goiter is most frequently made in the fifth or sixth decade of life, with a female/male rate of 4:1. retrosternal goiters can be classified as either primary or secondary. Primary intrathoracic goiters arise from aberrant thyroid tissue which is ectopically located in the mediastinum, receive their blood supply from mediastinal vessels and are not connected to the cervical thyroid. They are rare, representing less than 1% of all retrosternal goiters. (*Wu et al., 2006*).

Secondary retrosternal goiters develop from the thyroid located in its normal cervical site. Downward migration of the thyroid into the mediastinum is facilitated by negative intrathoracic pressure, gravity, traction forces during swallowing and the presence of anatomical barriers preventing the enlargement in other directions (thyroid cartilage, vertebral bodies, and strap muscles, especially in patients with a short neck). These secondary retrosternal goiters are, characteristically, in continuity with the cervical portion of the gland and receive their blood supply, depending on cervical vessels, almost always through branches of the inferior thyroid artery. retrosternal goiters show, in most cases, a slow growing enlargement, which usually remain asymptomatic for many years; about 20-40% of retrosternal goiters are discovered as an incidental finding on a radiographic examination. (*Shen et al., 2004*).

The most common symptoms are related to compression of the airways and the esophagus, and are represented by dyspnoea, choking, inability to sleep comfortably, dysphagia and hoarseness. Less commonly, signs of compression of vascular and nervous structures are present, such as superior vena cava obstruction (superior vena cava syndrome) and Horner's syndrome (compression of sympathetic chain). (*Mackle et al., 2007*).

The diagnosis of retrosternal goiter is based upon clinical history, clinical examinations, and imaging findings (*Mackle et al., 2007*).

Computed tomography (CT) scanning is, at present the most exhaustive examination for assessment of the extent of the goiter and compression effects on adjacent anatomical structures. (*Grainger et al., 2005*).

Magnetic resonance imaging (MRI) adds little additional information to that obtained with CT and is not routinely used. (*Page and Strunski, 2007*).

There is general agreement that surgical removal is the treatment of choice for retrosternal goiter, even in the absence of clinical symptoms. There are numerous reasons for performing surgery in such cases: non-surgical treatment of retrosternal goiter with thyroid hormone or radioactive iodine ablation is very rarely successful (*Hedayati and McHenry, 2002*); retrosternal goiter can become a life-threatening emergency if there is a sudden enlargement of the goitre, secondary to haemorrhage or malignant change; a diagnosis of malignancy, reported in 3-21% of retrosternal goiters (*Nervi et al., 1998*), could be missed, considering the difficulties and potential dangers in performing fine-needle aspiration cytology in the mediastinal portion of a retrosternal goiter.

Most retrosternal goiters can be removed through a cervical approach, while a partial or total sternotomy should be performed only in a minority of patients, ranging between 1-11% (*White et al., 2008*).

A careless attempt to remove a huge RSG using the cervical approach can lead to recurrent laryngeal nerve injury, which has been consistently reported after the surgery. There is an alternative and less invasive technique combining video-assisted thoracoscopic surgery (VATS) with a supraclavicular approach. This technique seems to offer improved exposure and reliable control of the neuro-vascular structures in the anterior mediastinum when resecting a huge substernal goiter that may prevent nerve injury (*Shigemura et al., 2005*).

There is also the robotic approach which By virtue of 3 dimensional visualization, greater dexterity, and more accurate dissection, the Da Vinci robot, for the first time, enables a completely minimally invasive approach to the posterior superior mediastinum, Robotic surgical techniques for removal of a retrosternal goiter and other thyroid masses with mediastinal extension, in combination with cervical incision, are effective. Robotic-assisted techniques can complement video-assisted thoracic surgical techniques and broaden the indications for minimally invasive surgery (*Whitson et al., 2007*).

Aim of the Work

This work aims at evaluating different types of retrosternal goiter with light focused on diagnosis, complication and recent trends in their management.

Thyroid Anatomy

Embryological development

In the fetus at 3-4 weeks of gestation, the thyroid gland appears as an epithelial proliferation in the floor of the pharynx at the base of the tongue between the tuberculum impar and the copula linguae at a point later indicated by the foramen cecum. The thyroid then descends in front of the pharyngeal gut as a bilobed diverticulum through the thyroglossal duct. Over the next few weeks, it migrates to the base of the neck, passing anterior to the hyoid bone. During migration, the thyroid remains connected to the tongue by a narrow canal, the thyroglossal duct. (*Berbel et al., 2010*).

Thyrotropin-releasing hormone (TRH) and thyroid-stimulating hormone (TSH) start being secreted from the fetal hypothalamus and pituitary at 18-20 weeks of gestation, and fetal production of thyroxine (T4) reach a clinically significant level at 18-20 weeks, fetal triiodothyronine (T3) remains low (less than 15 ng/dL) until 30 weeks of gestation, and increases to 50 ng/dL at term. (*Eugster et al., 2007*).

Fetal self-sufficiency of thyroid hormones protects the fetus against e.g. brain development abnormalities caused by maternal hypothyroidism. (*Zoeller et al., 2003*).

However, preterm births can suffer neurodevelopmental disorders due to lack of maternal thyroid hormones due to their own thyroid being insufficiently developed to meet their postnatal needs, the portion of the thyroid containing the parafollicular C cells, those responsible for the production of calcitonin, are derived from the neural crest. This is first seen as the ultimobranchial body, which joins the primordial thyroid gland during its descent to its final location in the anterior neck. (*Berbel et al., 2010*).

Over view:

The thyroid gland is a butterfly shaped, highly vascular, brownish red gland located anteriorly in the lower neck, extending from the level of the fifth cervical vertebra down to the first thoracic. The gland varies from an H to a U shape and is formed by 2 elongated lateral lobes with superior and inferior poles connected by a median isthmus, with an average height of 12-15mm, overlying the second to fourth tracheal rings. (See figure No 1) The isthmus is encountered during routine tracheotomy and must be retracted (superiorly or

inferiorly) or divided. Occasionally, the isthmus is absent, and the gland exists as 2 distinct lobes.(*Cummings et al., 1998*).

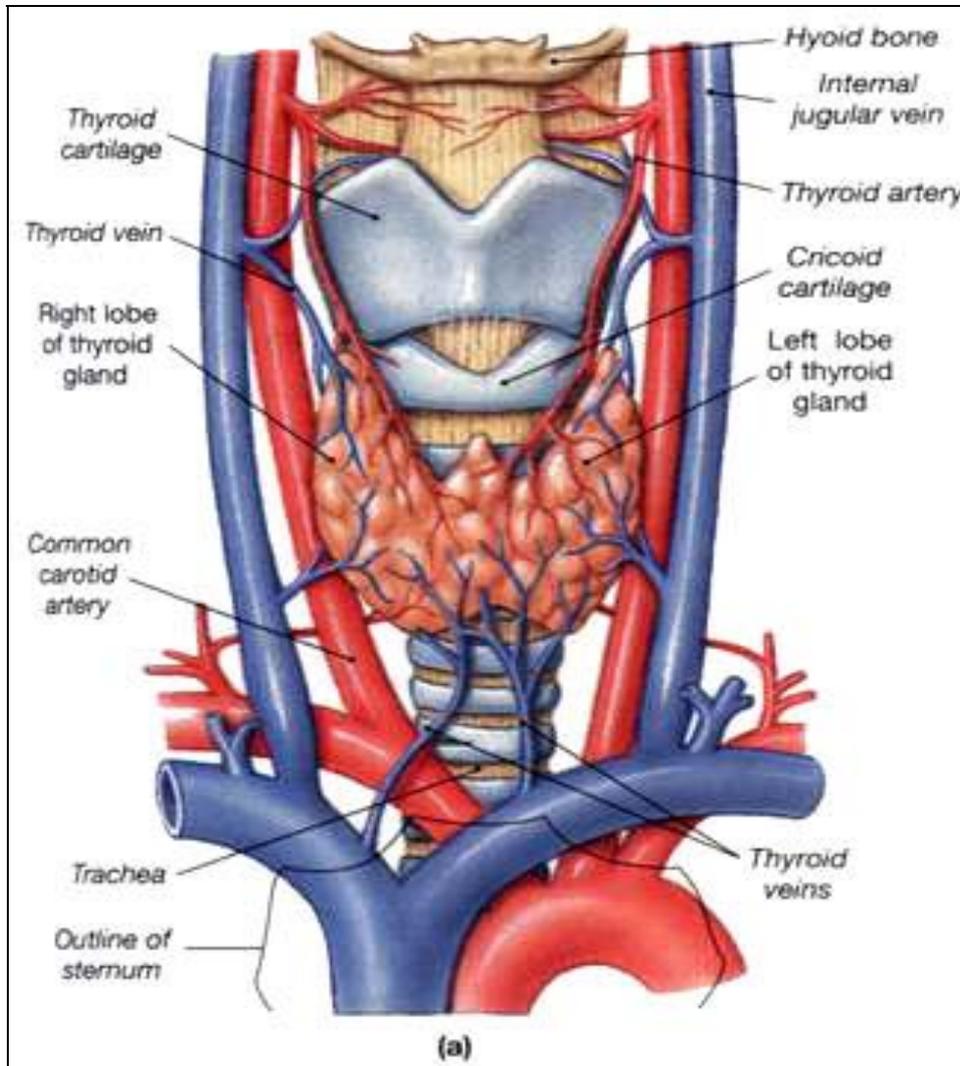


Fig. (1) Thyroid gland (*Lemaire and David, 2005*).