

Cancer Association of South Africa (CANSA)



Research • Educate • Support

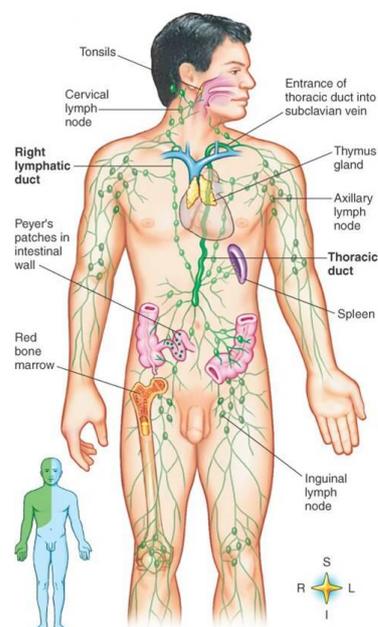
Fact Sheet on Thymoma

Introduction

The lymph system is made up of thin tubes that branch out to all parts of the body. The lymph system carries lymph, a colourless fluid containing a type of white blood cell called lymphocytes. Lymphocytes fight germs in the body. B-lymphocytes, or B cells, make antibodies to fight bacteria, and T-lymphocytes, or T cells, destroy viruses and foreign cells and trigger the B cells to make antibodies. The thymus is involved in the development of T-lymphocytes.

[Picture Credit: Lymphatic System]

Tiny, bean-shaped organs called lymph nodes are located throughout the body at different sites. Lymph nodes are found in clusters in the abdomen, groin, pelvis, underarms, and neck. In addition to the thymus, other parts of the lymph system include the spleen, which makes lymphocytes and filters blood, and the tonsils, located in the throat.



Cancer begins when normal cells change and grow uncontrollably, forming a mass called a tumour. A tumour can be cancerous or benign. A cancerous tumour is malignant, meaning it can spread to other parts of the body. A benign tumour means the tumour will not spread.

The Thymus Gland

The thymus gland will not function throughout an individual's full lifetime, but it has a big responsibility when it is active - helping the body protect itself against autoimmunity, which occurs when the immune system turns against itself. Therefore, the thymus plays a vital role in the lymphatic system (the body's defense network *and* endocrine system).

Researched and Authored by Prof Michael C Herbst

[D Litt et Phil (Health Studies); D N Ed; M Art et Scien; B A Cur; Dip Occupational Health; Dip Genetic Counselling; Dip Audiometry and Noise Measurement; Diagnostic Radiographer]

Reviewed by

Approved by Ms Elize Joubert, Chief Executive Officer [BA Social Work (cum laude); MA Social Work]

May 2021

The thymus is instrumental in the production and maturation of T-lymphocytes or T cells, a specific type of white blood cell that protects the body from certain threats, including viruses and infections. The thymus produces and secretes thymosin, a hormone necessary for T cell development and production.

[Picture Credit: Thymus Gland]



The thymus is special in that, unlike most organs, it is at its largest in children. Once one reaches puberty, the thymus starts to slowly shrink and become replaced by fat. By age 75, the thymus is little more than fatty tissue. Fortunately, the thymus produces all of one's T cells by the time one reaches puberty.

Thymosin: The Hormone of the Thymus - thymosin stimulates the development of T cells. Throughout childhood years, white blood cells called lymphocytes pass through the thymus, where they are transformed into T cells.

Thymoma (Thymus Cancer)

Thymoma is cancer that develops in the thymus gland. It is the uncontrollable growth of cells that eventually forms a tumour.

Robinson, S.P. & Akhondi, H. 2021.

“Thymomas and thymic carcinomas originate from the epithelial cells of the thymus within the anterior mediastinum. The thymus is an encapsulated bilobed gland. Each lobe of the thymus has a superior and inferior horn and extends laterally to each respective phrenic nerve. The thymus is supplied by branches off of the internal mammary, inferior thyroid, and pericardiophrenic arteries. It is drained by tributaries from the innominate vein or directly into the superior vena cava (SVC). The thymus is vital in the development of the adaptive immune system and commonly involutes after puberty into fibrofatty tissue. There has been a strong association between myasthenia gravis and thymomas that was found incidentally in 1939 by Alfred Blalock. The presence of histologic heterogeneity is common among thymomas, and as a result, benign and malignant thymomas cannot simply be differentiated based on history; however, malignant thymomas are much more invasive compared to their benign counterpart. The 15-year survival rate is 12.5% in patients with invasive thymomas and 47% in patients with noninvasive thymomas. Deaths related to thymomas usually occur from cardiac tamponade or other cardiorespiratory complications.”

Jilani, T.N. & Siddiqui, A.H. 2020.

“The mediastinum is a cavity that separates the lungs from the other structures in the chest. Generally, it is further divided into three main parts: anterior mediastinum, posterior mediastinum, and middle mediastinum. The borders of the mediastinum include the thoracic inlet superiorly, the diaphragm inferiorly, the spine posteriorly, the sternum anteriorly, and the pleural spaces laterally. Structures contained within the mediastinal cavity include the heart, aorta, esophagus, thymus, and trachea. Cancers in the mediastinum can develop from structures that are anatomically located

inside the mediastinum or that transverse through the mediastinum during development, and also from metastases or malignancies that originate elsewhere in the body.”

Causes of Thymoma

The exact cause of thymomas is not known. Thymomas are said to be slightly more common in men than in women and are most frequently seen in persons between the ages of 40 and 60. There are no known risk factors that predispose a person to developing thymoma.

Rich, J.T. 2020.

“Thymic tumours are a heterogeneous group of malignancies with a range of clinical presentations. The most common types are thymoma and thymic carcinoma, but overall it remains a rare cancer, and one without a clear aetiology. In this review of the epidemiology of the disease, the relationship between sex, age, and ethnicity is reviewed, along with limited evidence on the genetics of the condition. In terms of risk factors and potential causative factors, environmental exposures such as tobacco, radiation, alcohol, or diet, seem to be irrelevant, but there is some evidence linking the development of thymoma and thymic carcinoma with viral conditions, including Epstein Barr Virus. But data is not conclusive, and in the absence of large patient numbers, it is difficult to prove causation. There has been good research looking at the link between thymoma and other malignancies, either before or after the diagnosis. There does not appear to be a significant increased likelihood of thymoma following other malignancies. But, there is a suggestion, in several papers, that there is an increased risk of other malignancies following the diagnosis of thymoma, although the magnitude of this risk is disputed. There does appear to be an increased risk of non-Hodgkins Lymphoma after a diagnosis of thymoma, and this could be related to a disruption in T-cell function caused by either the disease process or the treatment directed at the thymoma. In summary though, it is a rare malignant process with a variety of presentations, often limited to the anterior mediastinum, and without an aggressive disease profile.”

The Incidence of Thymoma in South Africa

The incidence of thymoma in South Africa is not known as the National Cancer Registry (2017) does not make mention of thymoma.

Signs and Symptoms of Thymoma

Up to 50% of thymomas are asymptomatic, meaning they do not produce any symptoms or signs and are diagnosed during an imaging study for another reason. In other cases, the tumour may cause symptoms related to the size of the tumour and the pressure it exerts on adjacent organs:

- Chest pain
- shortness of breath
- cough
- Fever
- Night Sweats
- Weight loss

Researched and Authored by Prof Michael C Herbst

[D Litt et Phil (Health Studies); D N Ed; M Art et Scien; B A Cur; Dip Occupational Health; Dip Genetic Counselling; Dip Audiometry and Noise Measurement; Diagnostic Radiographer]

Reviewed by

Approved by Ms Elize Joubert, Chief Executive Officer [BA Social Work (cum laude); MA Social Work]

May 2021

Thymic Carcinoma

Thymic carcinomas are more aggressive tumours than thymomas and are more likely to spread and to cause symptoms. Thymic carcinoma is a much rarer condition than thymoma. It tends to grow and develop more quickly and is more likely to spread to other parts of the body. Thymic carcinomas are found in all age groups but are very rare. Most people do have symptoms. These include a cough and chest pain.

Diagnosis of Thymoma

Diagnosis of thymoma includes:

- a physical examination
- chest X-ray
- other imaging tests such as positron emission tomography (PET) scan, computed tomography (CT) scan, or magnetic resonance imaging (MRI)
- biopsy

Staging of Thymoma

The Masaoka-Koga Staging System is mostly used to stage thymomas.

Treatment of Thymoma

Treatment of thymoma includes one or more of the following:

- Surgery – removal of the tumour
- Chemotherapy
- Radiation therapy
- Hormone therapy

Jakopovic, M., Bitar, L., Seiwerth, F., Marusic, A., Krpina, K. & Samarzija, M. 2020.

“Thymic epithelial tumors (TETs) are rare thymic neoplasms. There are approximately 1.5 cases per million TETs per year. They are the most common anterior mediastinal tumors in adults. Due to limited activity of available treatment options novel strategies and treatment options are needed and treatment with immune checkpoint inhibitors is an attractive option. Thymic epithelial tumors have one of the lowest tumor mutational burden among all cancer in adults, but high expression of PD-L1 on tumor cells and abundant CD8+ lymphocytes provide a strong rationale for implementing immune checkpoint inhibitors (ICIs) which target PD-1/PD-L1 pathway in the treatment of TETs. Few small early stage clinical trials were published so far evaluating efficacy of pembrolizumab and avelumab in thymoma and thymic carcinoma patients. All trials showed reasonable response rates and progression-free survival. Higher PD-L1 expression was predictor of response in all trials. However, increased incidence of immune-related adverse events was seen in TET patients treated with immune checkpoint inhibitors compared to patients with other cancers. At the moment, ICIs

Researched and Authored by Prof Michael C Herbst

[D Litt et Phil (Health Studies); D N Ed; M Art et Scien; B A Cur; Dip Occupational Health; Dip Genetic Counselling; Dip Audiometry and Noise Measurement; Diagnostic Radiographer]

Reviewed by

Approved by Ms Elize Joubert, Chief Executive Officer [BA Social Work (cum laude); MA Social Work]

May 2021

are not standard of care for patients with TET and larger trials are needed to establish the right role of ICIs regarding efficacy and safety of these agents.”

Terra, R.M., Milanez-de-Campos, J.R., Haddad, R., Tridade, J.R.M., Lauricella, L.L., Ribas, B.J. & Pêgo-Fernandes, P.M. 2019.

Objective: To evaluate the results of resection of tumors of the thymus by robotic thoracic surgery, analyzing the extent of resection, postoperative complications, time of surgery, and length of stay.

Methods: Retrospective study from a database involving patients diagnosed with a tumor of the thymus and undergoing robotic thoracic surgery at one of seven hospitals in Brazil between October of 2015 and June of 2018.

Results: During the study period, there were 18 cases of resection of tumors of the thymus: thymoma, in 12; carcinoma, in 2; and carcinoid tumor, in 1; high-grade sarcoma, in 1; teratoma, in 1; and thymolipoma, in 1. The mean lesion size was 60.1 ± 32.0 mm. Tumors of the thymus were resected with tumor-free margins in 17 cases. The median (interquartile range) for pleural drain time and hospital stay, in days, was 1 (1-3) and 2 (2-4), respectively. There was no need for surgical conversion, and there were no major complications.

Conclusions: Robotic thoracic surgery for resection of tumors of the thymus has been shown to be feasible and safe, with a low risk of complications and with postoperative outcomes comparable to those of other techniques.

About Clinical Trials

Clinical trials are research studies that involve people. They are conducted under controlled conditions. Only about 10% of all drugs started in human clinical trials become an approved drug.

Clinical trials include:

- Trials to test effectiveness of new treatments
- Trials to test new ways of using current treatments
- Tests new interventions that may lower the risk of developing certain types of cancers
- Tests to find new ways of screening for cancer

The [South African National Clinical Trials Register](#) provides the public with updated information on clinical trials on human participants being conducted in South Africa. The Register provides information on the purpose of the clinical trial; who can participate, where the trial is located, and contact details.

For additional information, please visit: www.sanctr.gov.za/

Medical Disclaimer

This Fact Sheet is intended to provide general information only and, as such, should not be considered as a substitute for advice, medically or otherwise, covering any specific situation. Users should seek appropriate advice before taking or refraining from taking any action in reliance on any information contained in this Fact Sheet. So far as permissible by law, the Cancer Association of South Africa (CANSA) does not accept any liability to any person (or his/her dependants/estate/heirs) relating to the use of any information contained in this Fact Sheet.

Researched and Authored by Prof Michael C Herbst

[D Litt et Phil (Health Studies); D N Ed; M Art et Scien; B A Cur; Dip Occupational Health; Dip Genetic Counselling; Dip Audiometry and Noise Measurement; Diagnostic Radiographer]

Reviewed by

Approved by Ms Elize Joubert, Chief Executive Officer [BA Social Work (cum laude); MA Social Work]

May 2021

Whilst the Cancer Association of South Africa (CANSA) has taken every precaution in compiling this Fact Sheet, neither it, nor any contributor(s) to this Fact Sheet can be held responsible for any action (or the lack thereof) taken by any person or organisation wherever they shall be based, as a result, direct or otherwise, of information contained in, or accessed through, this Fact Sheet.



Sources and References Consulted or Utilised

Comacchio, G.M., Marulli, G., Mammana, M., Natale, G., Schiavon, M. & Rea, F. 2019. Surgical decision making: thymoma and Myasthenia Gravis. *Thorac Surg Clin*, 29 (2), 203-213. May 2019

Gorospe, S.L., Almeida-Aróstegui, N.A. & Arrieta, P. 2018. Core needle biopsy of an anterior mediastinal thymoma: creation of a safe access route by hydrodissection. *Arch Bronconeumol*. 2018 Jan 17. pii: S0300-2896(17)30435-0. doi: 10.1016/j.arbres.2017.12.001. [Epub ahead of print] English, Spanish. No abstract available. PMID: 29373145.

Helm, J.M., Lavy, D., Figueroa-Bodine, J. & Joseph, S. 2017. Metastatic malignant thymoma to the abdomen: a SEER database review and assessment of treatment strategies. *World J Oncol*. 2017 Oct;8(5):147-150. doi: 10.14740/wjon1057w. Epub 2017 Nov 5. PMID: 29147451.

Hor, J.Y., Lim, T.T., Cheng, M.C., Chia, Y.K., Wong, C.K., Lim, S.M., Cheah, C.F., Tan, K., Easawm P.E.S. & Leite, M.I. 2018. Thymoma-associated myasthenia gravis and LHI1-encephalitis, with nephrotic syndrome post-thymectomy. *J Neuroimmunol*. 2018 Jan 30. pii: S0165-5728(17)30491-5. doi: 10.1016/j.jneuroim.2018.01.011. [Epub ahead of print]. PMID: 29395322.

Jakopovic, M., Bitar, L., Seiwert, F., Marusic, A., Krpina, K. & Samarzija, M. 2020. Immunotherapy for thymoma. *J Thorac Dis*. 2020 Dec;12(12):7635-7641.

Jilani, T.N. & Siddiqui, A.H. 2020. Mediastinal Cancer. In: *StatPearls [Internet]*. Treasure Island (FL): StatPearls Publishing; 2020 Jan-. 2020 Jan 15.

Kaba, E., Ozkan, B., Erus, S., Duman, S., Cimenoglu, B. & Toker, A. 2017. Role of surgery in the treatment of Masaoka stage Iva thymoma. *Ann Thorac Cardiovasc Surg*. 2017 Dec 8. doi: 10.5761/atcs.0a.17-00108. [Epub ahead of print]. PMID: 29225302.

Krishan, M. & Ganti, A.K. 2019. The role of targeted therapy in thymic carcinoma. *J Oncol Pharm Pract*, 25 (7), 1712-1718. Oct 2019.

Lymphatic System

https://www.google.co.za/search?q=lymphatic+system&source=lnms&tbm=isch&sa=X&ei=n6XYU4e4G9KA7QbFzIH4DA&sqi=2&ved=0CAYQ_AUoAQ&biw=1517&bih=714&dpr=0.9#facrc=_&imgdii=_&imgrc=YFSA26FR7t-uAM%253A%3BLbtzHbqOLOvJwM%3Bhttp%253A%252F%252Fhealthfavo.com%252Fwp-content%252Fuploads%252F2013%252F10%252Flymphatic-system-anatomy.jpg%3Bhttp%253A%252F%252Fhealthfavo.com%252Flymphatic-system-anatomy.html%3B528%3B865

Marulli, G., Comacchio, G.M. & Rea, F. 2017. Robotic thymectomy. *J Vis Surg*. 2017 May 11;3:68. doi: 10.21037/jovs.2017.05.01. eCollection 2017. Review. PMID: 29078631.

Mignarri, A., Gentili, F., Masia, F., Genua, A., Cenciarelli, S., Brunori, P., Mazzei, M.A., Malandrini, A., Federico, A., Mazzei, F.G. & Dotti, M.T. 2017. Imaging of the thymus in myotonic dystrophy type 1. *Neurol Sci*. 2017 Nov 25. doi: 10.1007/s10072-017-3202-4. [Epub ahead of print] PMID: 29177794.

Researched and Authored by Prof Michael C Herbst

[D Litt et Phil (Health Studies); D N Ed; M Art et Scien; B A Cur; Dip Occupational Health; Dip Genetic Counselling; Dip Audiometry and Noise Measurement; Diagnostic Radiographer]

Reviewed by

Approved by Ms Elize Joubert, Chief Executive Officer [BA Social Work (cum laude); MA Social Work]

May 2021

Odaka, M., Tsukamoto, Y., Shibasaki, T., Mori, S., Asano, H., Yamashita, M. & Morikawa, T. 2017. Surgical and oncological outcomes of thoracoscopic thymectomy for thymoma. *J Vis Surg.* 2017 Apr 10;3:54. doi: 10.21037/jovs.2017.03.18. eCollection 2017. Review. PMID: 29078617.

PDQ Adult Treatment Editorial Board. 2017. Thymoma and thymic carcinoma treatment (PDQ®): patient version. PDQ Cancer Information Summaries [Internet]. Bethesda (MD): National Cancer Institute (US); 2002-. 2017 Nov 9. PMID: 26389395.

Rich, J.T. 2020. Epidemiology of thymoma. *J Thorac Dis.* 2020 Dec;12(12):7531-7535.

Robinson, S.P. & Akhondi, H. 2021. Thymoma. *In: StatPearls* [Internet]. Treasure Island (FL): StatPearls Publishing; 2021 Jan. 2021 Apr 7.

Tassi, V., Ceccarelli, S., Zannori, C., Gili, A., Daddi, N., Bellezza, G., Ascani, S., Liberati, A.M. & Puma, F. 2017. Could thymectomy be a reasonable option for non-myasthenic thymoma patients. *J Thorac Dis.* 2017 Oct;9(10):3817-3824. doi: 10.21037/jtd.2017.09.109. PMID: 29268390.

Terra, R.M., Milanez-de-Campos, J.R., Haddad, R., Tridade, J.R.M., Lauricella, L.L., Ribas, B.J. & Pêgo-Fernandes, P.M. 2019. Robotic thoracic surgery for resection of thymoma and tumors of the thymus: technical development and initial experience. *J Bras Pneumol*, 46 (1), e20180315. 2019 Dec 13 eCollection 2020.

The Oncologist. Thymoma: benign appearance, malignant potential.
<http://theoncologist.alphamedpress.org/content/11/8/887.full>.

Thymus Gland

https://www.google.co.za/search?q=thymoma+cancer&source=Inms&tbm=isch&sa=X&ei=yqPYU5rAF4yh7AampYHQDQ&sqi=2&ved=0CAYQ_AUoAQ&biw=1517&bih=714&dpr=0.9#facrc=_&imgdii=_&imgrc=yRIQIThbHV-9PM%253A%3BfB1kw6Ja7S1WqM%3Bhttp%253A%252F%252Fbionews-tx.com%252Fwp-content%252Fuploads%252F2013%252F09%252Fthymic-tumors.jpg%3Bhttp%253A%252F%252Fbionews-tx.com%252Fnews%252F2013%252F09%252F26%252Fcastle-biosciences-develops-genetic-test-that-accurately-differentiates-thymomas-from-thymic-carcinoma-tumors%252F%3B400%3B300

Researched and Authored by Prof Michael C Herbst

[D Litt et Phil (Health Studies); D N Ed; M Art et Scien; B A Cur; Dip Occupational Health; Dip Genetic Counselling; Dip Audiometry and Noise Measurement; Diagnostic Radiographer]

Reviewed by

Approved by Ms Elize Joubert, Chief Executive Officer [BA Social Work (cum laude); MA Social Work]

May 2021