# Mediastinal Disorders

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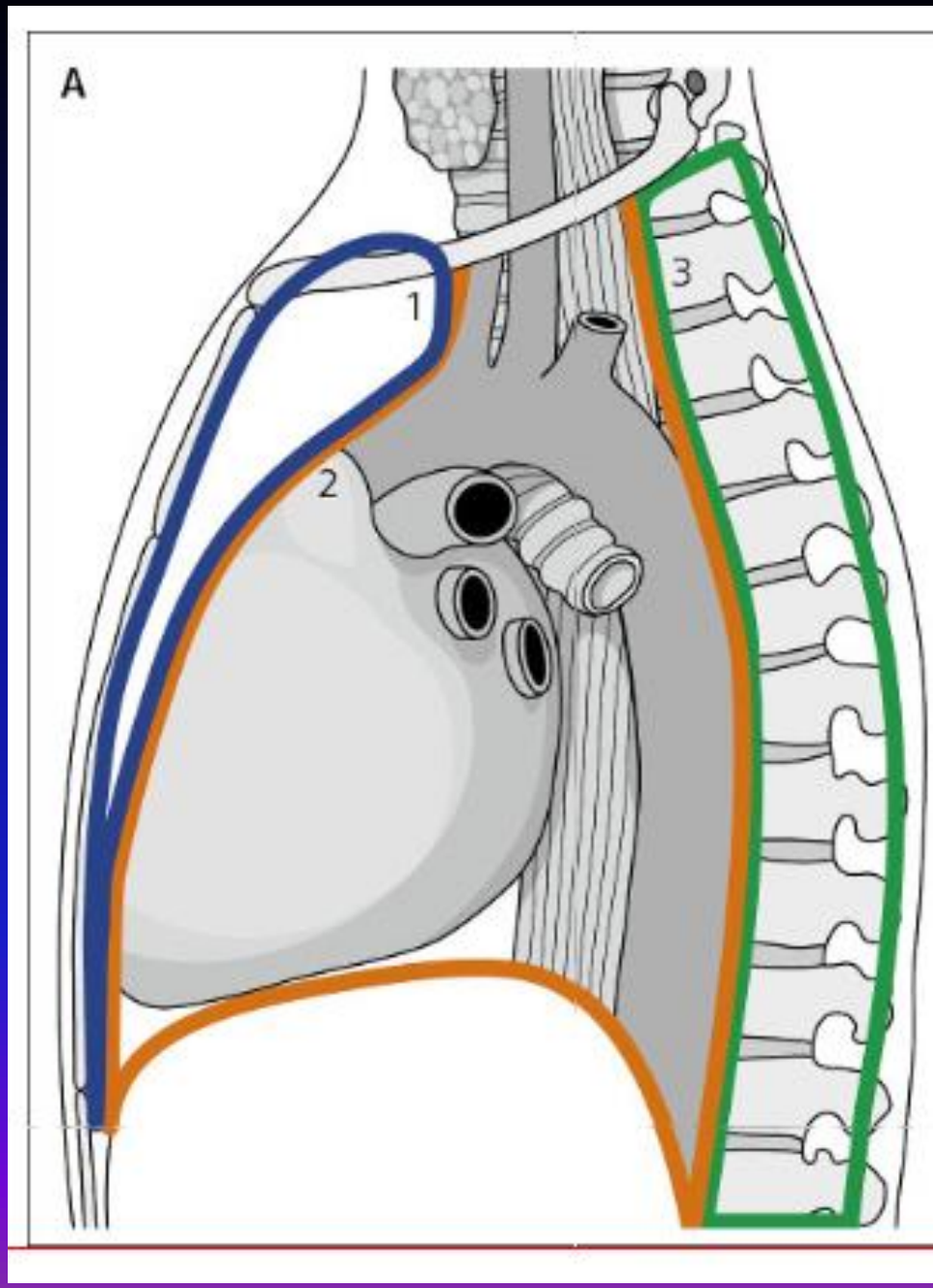
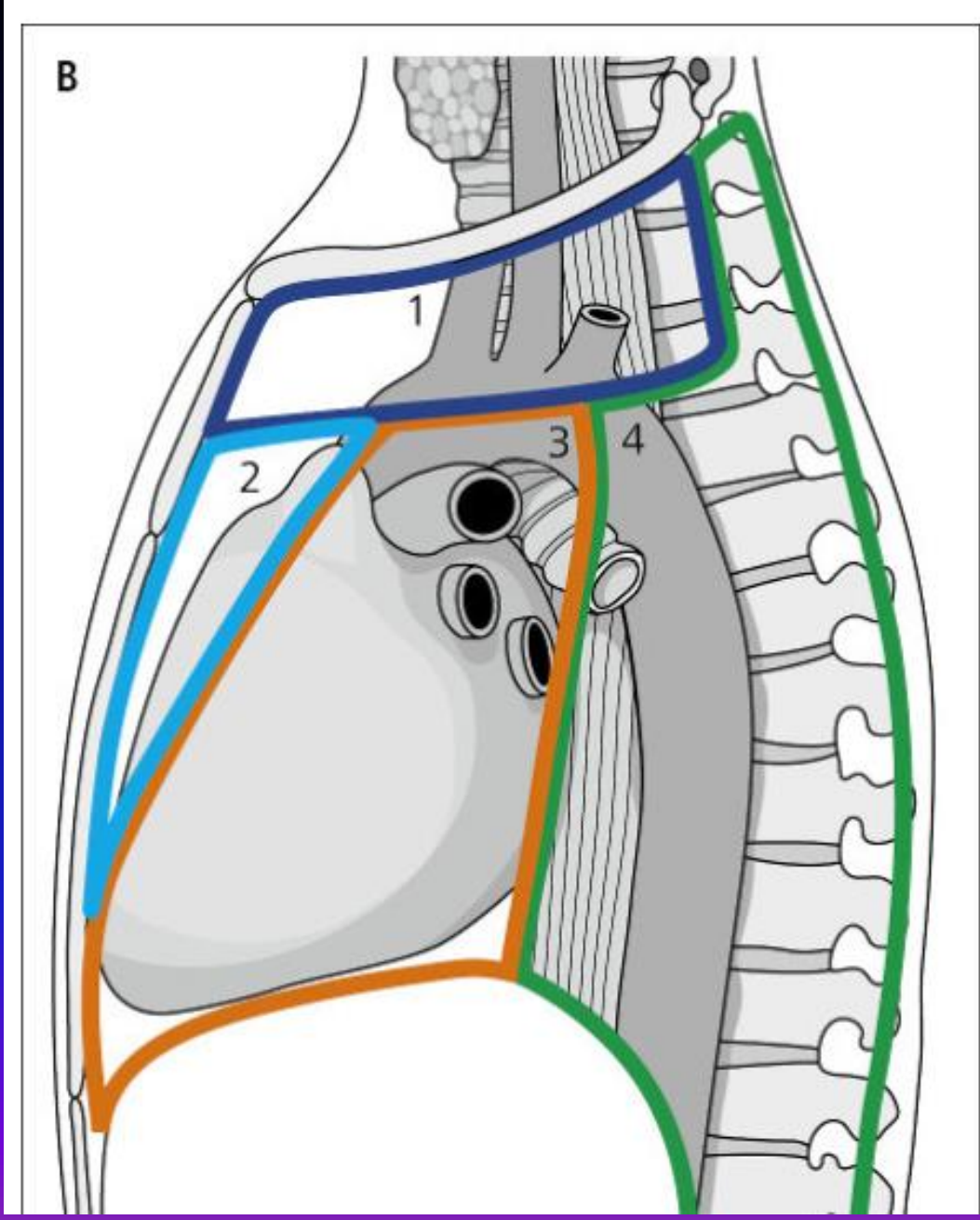
# Anatomy

- The *mediastinum* is anatomically defined as the space between the two lungs; it is demarcated by the thoracic inlet superiorly, the diaphragm inferiorly, the sternum anteriorly, and by the spine and paravertebral thoracic sulci posteriorly.
- For practical reasons, it is further divided into anterior, middle, and posterior compartments based on anatomic landmarks seen on lateral radiographs.
- Because the localization, extent, and radiologic characteristics of a mediastinal mass are best evaluated on (CT) scans, these landmarks are applied there accordingly.

- Shields' Three-Zone Model
  - Anterior
  - Visceral
  - paravertebral



- Traditional four compartments model.
  - Superior
  - Anterior
  - Middle
  - Posterior



# Boundaries and contents of the 4-compartment scheme

| | Boundaries | Contents |
|---|---|---|
| **Superior mediastinum** | Thoracic inlet to line drawn from angle of Louis to T4 | Aorta and great vessels, trachea, upper third of esophagus, upper thymus |
| **Anterior mediastinum** | Anterior to pericardium and posterior to body of sternum | Mediastinal fat and thymus |
| **Middle mediastinum** | Bounded anteriorly and posteriorly by pericardium | Pericardium and contents, carina, lymph nodes |
| **Posterior mediastinum** | Dorsal surface of pericardium to anterior surfaces of T4-T12 | Esophagus, descending thoracic aorta, azygos vein, thoracic duct |

# Boundaries and contents of Shields' 3-zone classification

| | Boundaries | Contents |
|---|---|---|
| **Previsceral zone (anterior)** | Under surface of sternum to anterior surface of pericardium and great vessels | Thymus, internal mammary vessels |
| **Visceral zone** | Anterior reflection of pericardium to anterior surface to spine | Pericardium and contents, trachea and main bronchi, esophagus |
| **Retrovisceral zone** | Paravertebral sulci | Costovertebral junctions, proximal intercostal nerves and arteries, sympathetic trunks |

# Mediastinal Disorders

*Non-neoplastic conditions*

- Acute necrotizing mediastinitis
- Chronic mediastinitis
- Mediastinal cysts (acquired and congenital)
- Mediastinal hemorrhage
- Pericardial disease
  - Acute pericarditis
  - Constrictive pericarditis
  - Pericardial effusion and cardiac tamponade

*Neoplastic conditions(Tumors)*

- Benign
- Malignant

# Anatomical distribution of Mediastinal Masses

| | Adults | Children |
|---|---|---|
| Anterior mediastinum | = 54% | 43% |
| Middle mediastinum | = 20% | 18% |
| Posterior mediastinum | = 26% | 39% |

- predilection for malignancy:
  - Anterior mediastinum = 60%
  - Middle mediastinum = 30%
  - Posterior mediastinum = 15%

# Incidence

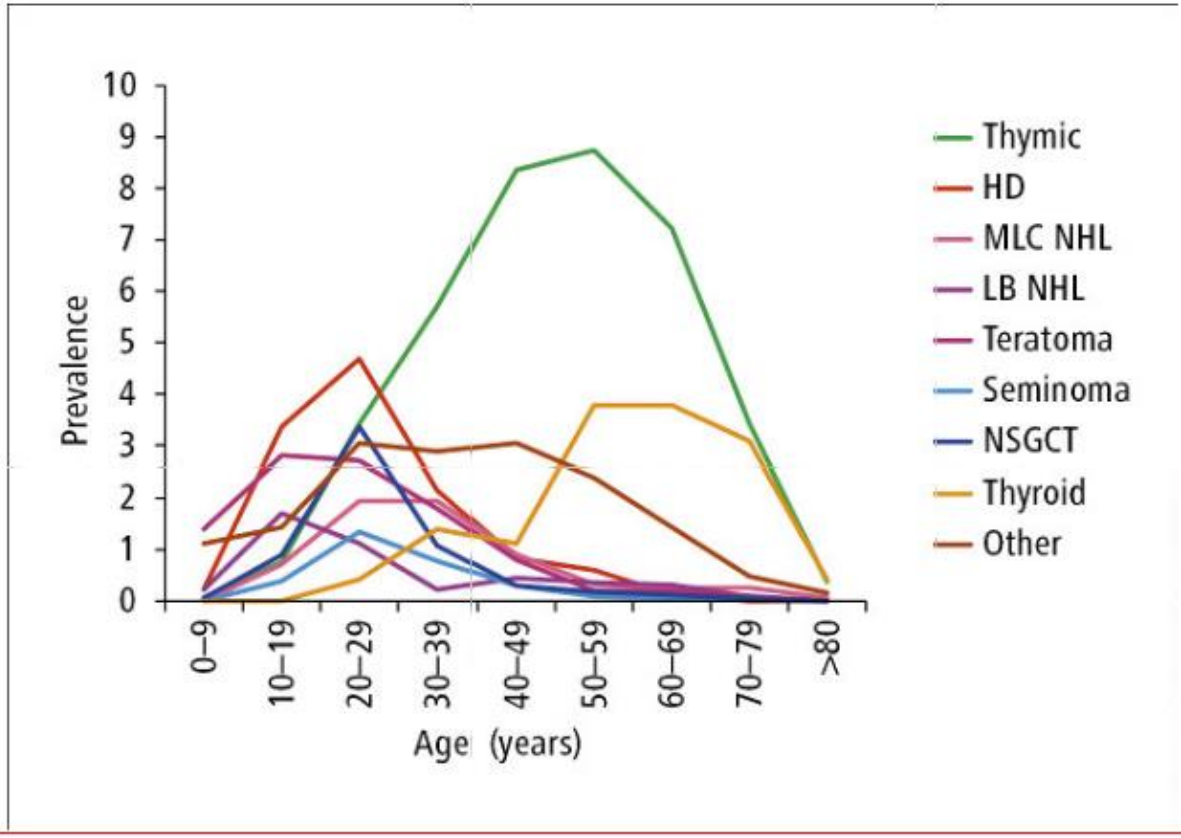



**Fig. 72-2.** Overall age-specific incidence of anterior mediastinal tumours (see Appendix). HD – Hodgkin disease; LB NHL – lymphoblastic non-Hodgkin lymphoma; MLC NHL – mediastinal large cell non-Hodgkin lymphoma; NSGCT – non-seminomatous germ cell tumor; teratoma – refers only to benign primary teratoma.

# Anterior Mediastinal Masses

- MC group of mediastinal tumors.
  - Thymoma 35%
    - MC in those above age of 40 year.
  - Lymphoma 25%

# Anterior / above 40

Thymoma

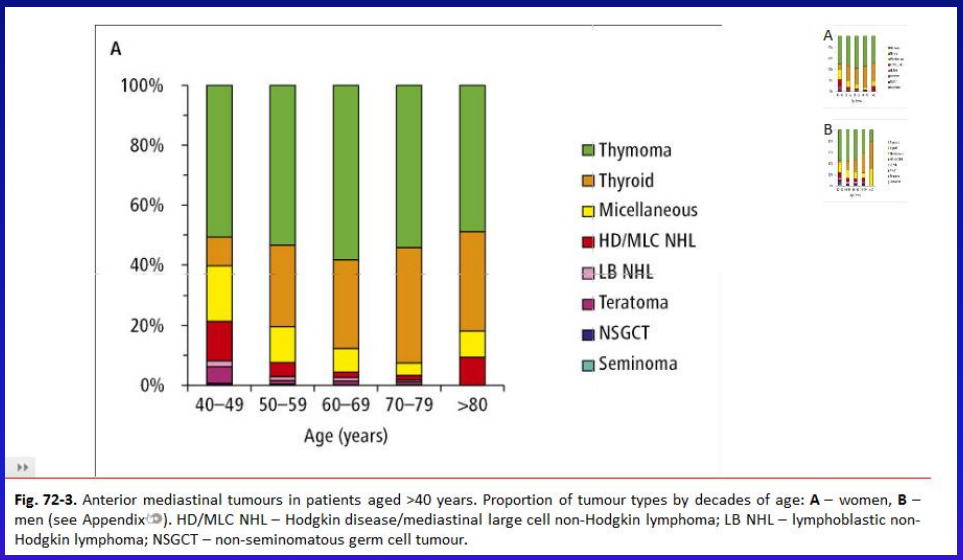
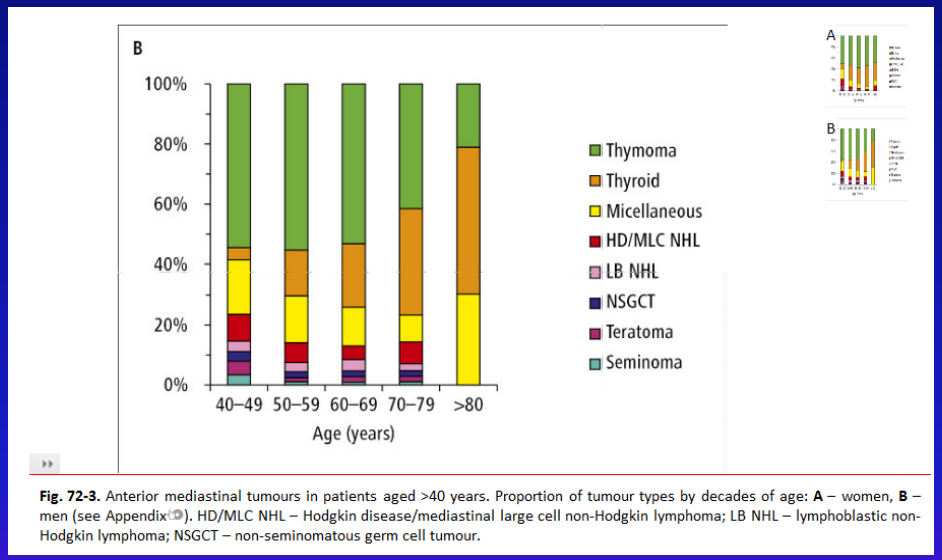



**Fig. 72-3.** Anterior mediastinal tumours in patients aged >40 years. Proportion of tumour types by decades of age: **A** – women, **B** – men (see Appendix). HD/MLC NHL – Hodgkin disease/mediastinal large cell non-Hodgkin lymphoma; LB NHL – lymphoblastic non-Hodgkin lymphoma; NSGCT – non-seminomatous germ cell tumour.

Thymoma

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# Anterior / 10-39 yr

Lymphoma

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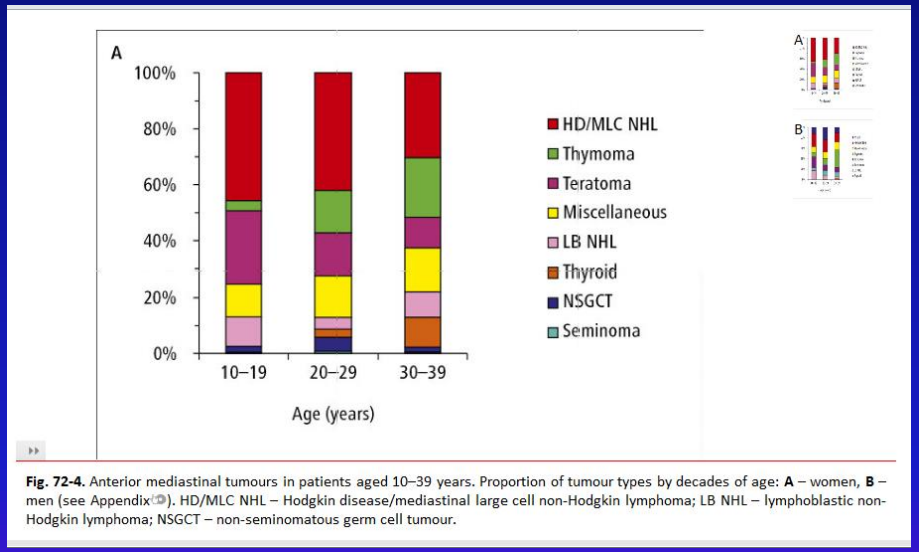
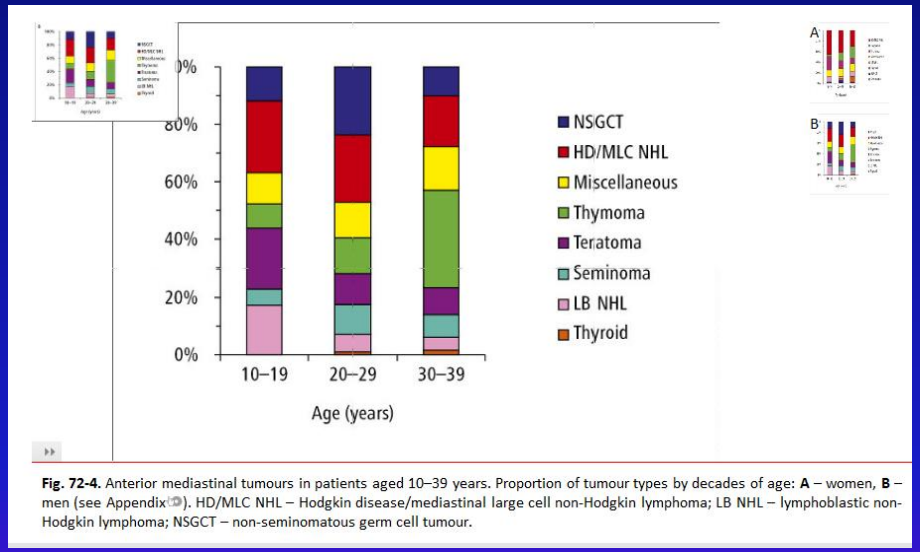



**Fig. 72-4.** Anterior mediastinal tumours in patients aged 10–39 years. Proportion of tumour types by decades of age: **A** – women, **B** – men (see Appendix). HD/MLC NHL – Hodgkin disease/mediastinal large cell non-Hodgkin lymphoma; LB NHL – lymphoblastic non-Hodgkin lymphoma; NSGCT – non-seminomatous germ cell tumour.

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# 0-9 yr

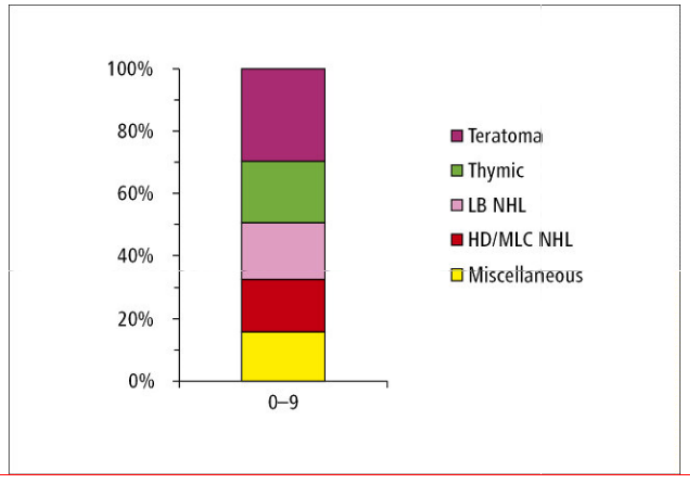
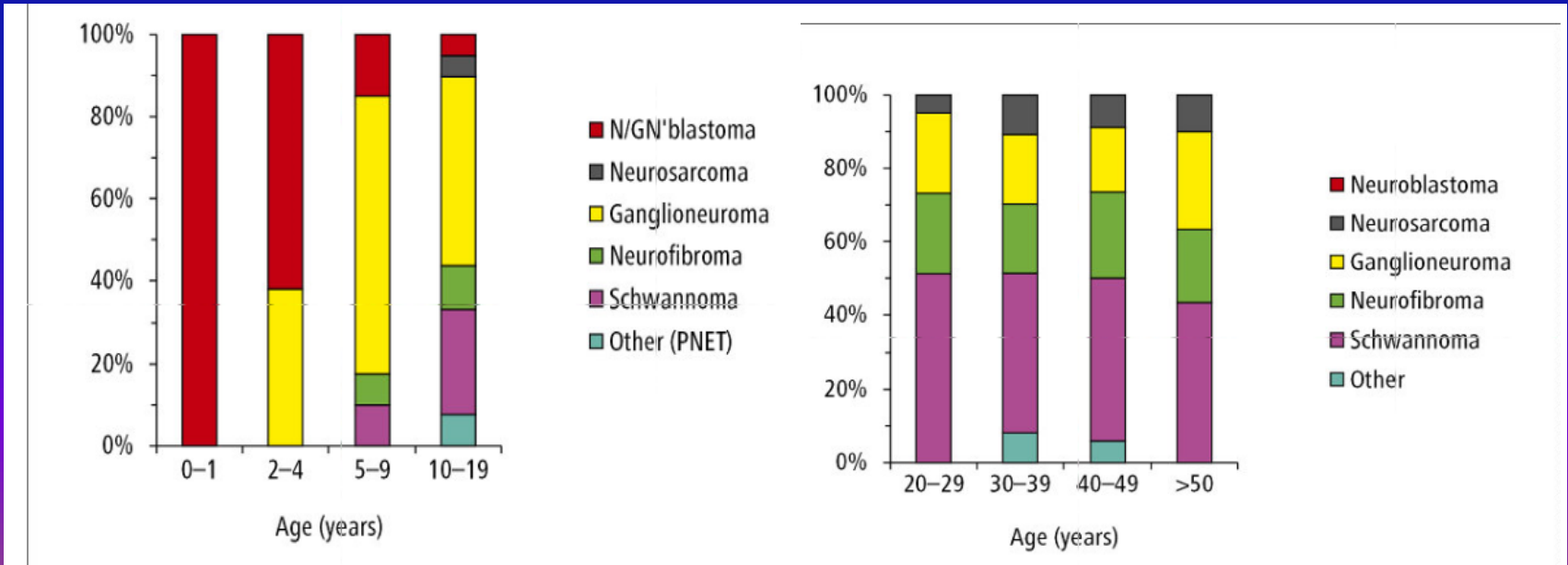



**Fig. 72-5.** Anterior mediastinal tumours in children aged <10 years. Proportion of tumour types by years of age (see Appendix ).

# Paravertebral Mediastinal Tumors

- In adults: MC benign nerve sheath tumors ( schwannoma, neurofibroma, ganglioneuroma)
- Children:
  - Less than 1 yr: malignant neuroblastoma or ganglioneuroblastoma



# Middle mediastinal Tumors

- Majority is benign. 50% are cysts.

# History and Physical Examination

- Age of the patient.
  - ***In adults,*** the most common tumors include neurogenic tumors of the posterior compartment, benign cysts occurring in any compartment, and thymomas of the anterior mediastinum .
  - ***In children***, neurogenic tumors of the posterior mediastinum are also common. Lymphoma is the second most common mediastinal tumor, usually located in the anterior or middle compartment, and thymoma is rare .
- In both age groups, approximately 25% of mediastinal tumors are malignant

# History and Physical Examination

- 2/3 in adults are discovered as asymptomatic.
- If symptomatic, these tumors are significantly more likely to be malignant.
- Size, location, rate of growth, and associated inflammation are important factors that correlate with symptoms.
- Large, bulky tumors, expanding cysts, and teratomas can cause compression, in particular the trachea, and lead to cough, dyspnea on exertion, or stridor.

# History and Physical Examination

- Chest pain or dyspnea may be reported secondary to associated pleural effusions, cardiac tamponade, or phrenic nerve involvement.
- Occasionally, hoarseness because of left recurrent laryngeal nerve involvement .
- Systemic Sx present in 50% of patients and a lymphoproliferative disorder, compared with only 29% of patients with other masses (such as thymic or neurogenic lesions).

# Key Points for Diagnostic Evaluation

- The location of mediastinal masses—anterior, middle, and posterior compartments—has important implications for diagnostic stragtegies and treatment
- Contrast enhanced CT is the most important imaging modality
- MRI is superior to CT in
  - ✓ Evaluation of tumor involvement of the great vessels and spine
  - ✓ Differentiating vascular, solid, and cystic lesions.
- FDG-PET imaging, or (PET-CT) imaging, is helpful in the staging of lymphoma and in the follow-up of patients with lymphoma and other malignant mediastinal tumors.

# Key Points for Diagnostic Evaluation

- Cytohistopathologic diagnosis is often required to confirm a presumed diagnosis based on clinical evaluation and on the radiologic characteristics on CT scan, especially for mediastinal masses located in the anterior mediastinum
- Techniques used for diagnosis and therapy of mediastinal masses are strongly related to their availability and to the expertise of the investigators.

# Key Points for Diagnostic Evaluation

- Cystic lesions and well-encapsulated mediastinal masses smaller than 7 cm can be completely removed (diagnostic and therapeutic)
- The recent introduction of new techniques and new instruments has changed the strategic decision about how to manage a mediastinal mass, allowing a more accurate diagnosis and the use of minimally invasive surgical procedures

# Diagnostic Evaluation

***1.Imaging***

- CXR = generally poorly defined.
- Contrast enhanced CT–chest = the most common imaging modality for evaluation of mediastinal masses.
- MRI = is more accurate than CT in determining if there is invasion of vascular structures or spinal involvement.

# Diagnostic Evaluation

- Single-photon emission computed tomography (SPECT).
- Iodine 131 or iodine 123 scan
- Octreotide scans or *m*-iodobenzylguanidine (iobenguane I 123, or MIBG).
- Sestamibi scan
- FDG-PET or FDG-PET-CT scanning

# Diagnostic Evaluation

## 2. Serum Markers

- Alpha-fetoprotein (AFP)
- Human chorionic gonadotropin (hCG).
  - In over 90% of nonseminomatous germ cell tumors, either the AFP or the hCG level will be elevated. Results are close to 100% specific if the level of either AFP or hCG is >500 ng/mL.
- Intact parathyroid hormone level for ectopic parathyroid adenomas.

# Diagnostic Evaluation

## 3. Nonsurgical and surgical biopsies:

- Percutaneous transthoracic biopsy
- EBUS + Biopsy
- EUS + Biopsy
- Video-Mediastinoscopy
- Anterior Mediastinotomy
- VATS
- Lateral Thoracotomy
- Sternotomy

# Anterior mediastinal masses

*Benign*

- Thymoma
- Thymic cyst
- Thymolipoma
- Thymic hyperplasia
- Thyroid
- Teratoma
- Cystic hygroma
- Parathyroid adenoma
- Foramen of morgagni hernia

*Malignant*

- Thymic carcinoma
- Thyroid carcinoma
- Seminoma
- Mixed germ cell
- Lymphoma
- Thymic carcinoid

# Thymoma

- Most common tumor of the anterior mediastinum.
- Usually in adults with a median age of 50 years with no gender preference.
- 40% of patients have paraneoplastic disorders such as MG (30%), pure red cell aplasia, or hypogammaglobulinemia (5%-10%).
- Others, such as SLE, Cushing's syndrome, and SIADH.

# Histological Classification

**BOX 44-2** World Health Organization Classification of Thymic Epithelial Tumors

| Type | Pathologic Classification | Prognosis |
|---|---|---|
| A | Medullary thymoma | Benign clinical course |
| | Spindle cell thymoma | |
| AB | Mixed thymoma | |
| B1 | Lymphocyte-rich thymoma | Moderately malignant clinical course |
| | Lymphocytic thymoma | |
| | Predominately cortical thymoma | |
| | Organoid thymoma | |
| B2 | Cortical thymoma | |
| B3 | Epithelial thymoma | |
| | Atypical thymoma | |
| | Squamoid thymoma | |
| | Well-differentiated thymic carcinoma | |
| C | Thymic carcinoma | Highly malignant clinical course |

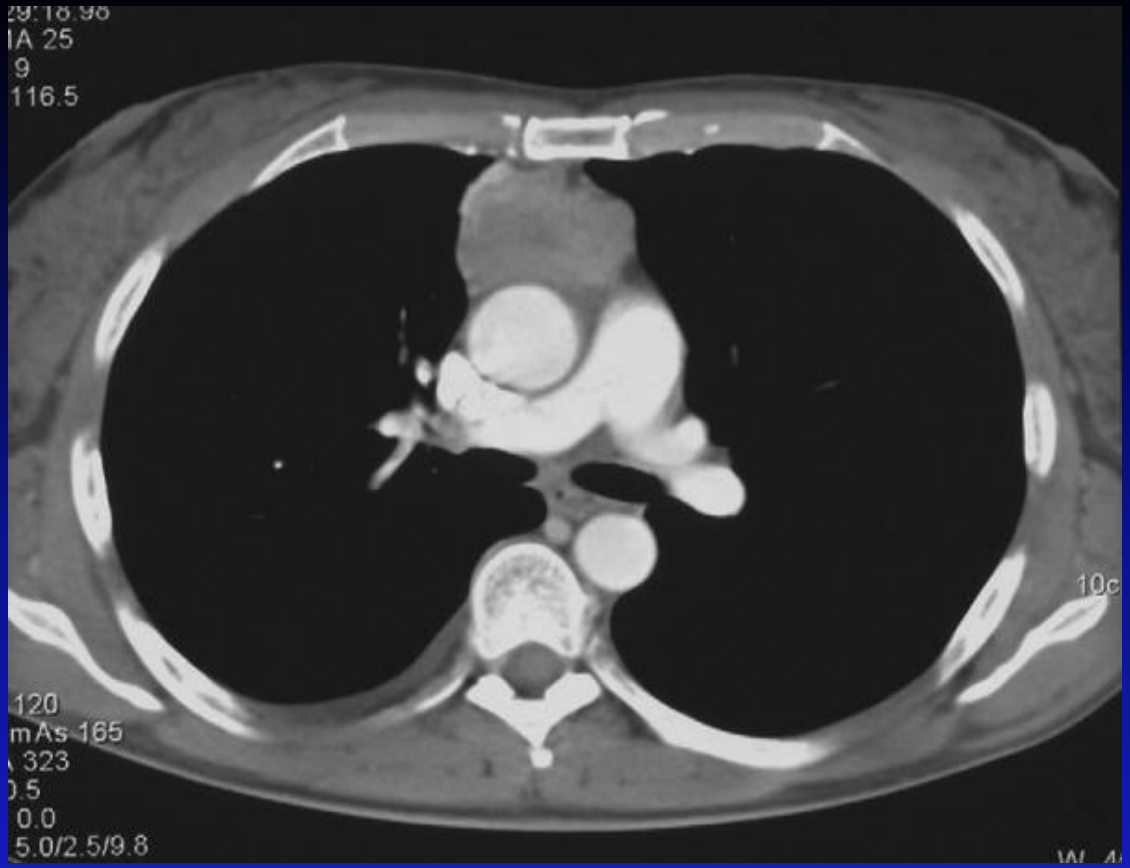
# Type A Thymoma

- Composed of spindle-shaped epithelial cells without atypia and No lymphocytes.
- Elderly
- 20% with MG
- Survival exceeds 80% at 5 and 10 years.

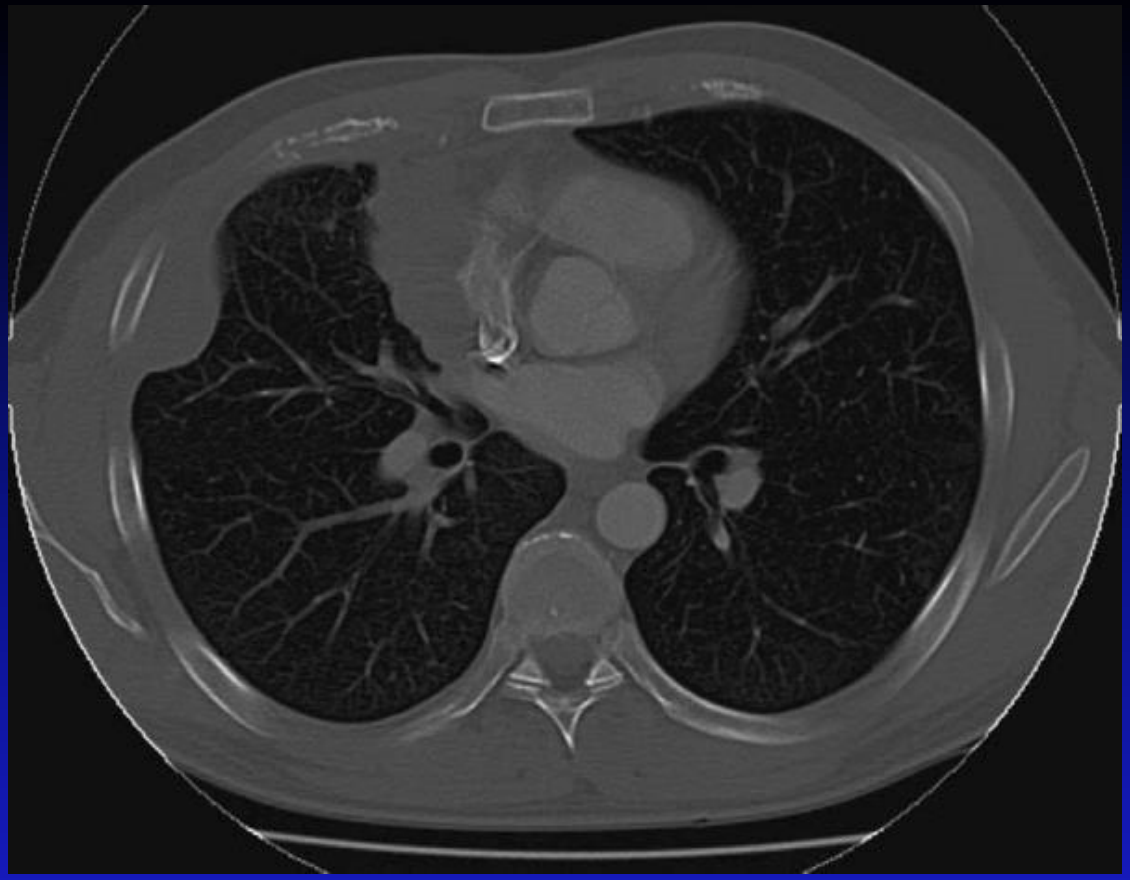
# Type B Thymoma

- 3 subtypes:
  - Subtype B1:
    - A tumor that resembles the normal functional thymus in that it combines large expanses having an appearance practically indistinguishable from normal thymic cortex with areas resembling thymic medulla
  - Subtype B2:
    - A tumor in which the neoplastic epithelial component appears as scattered plump cells with vesicular nuclei and distinct nucleoli among a heavy population of lymphocytes. Perivascular spaces are common and sometimes very prominent. A perivascular arrangement of tumor cells resulting in a palisading effect may be seen.

- Subtype B3:
  - A type of thymoma predominantly composed of epithelial cells having a round or polygonal shape and exhibiting no or mild atypia. They are admixed with a minor component of lymphocytes, resulting in a sheetlike growth of the neoplastic epithelial cells.



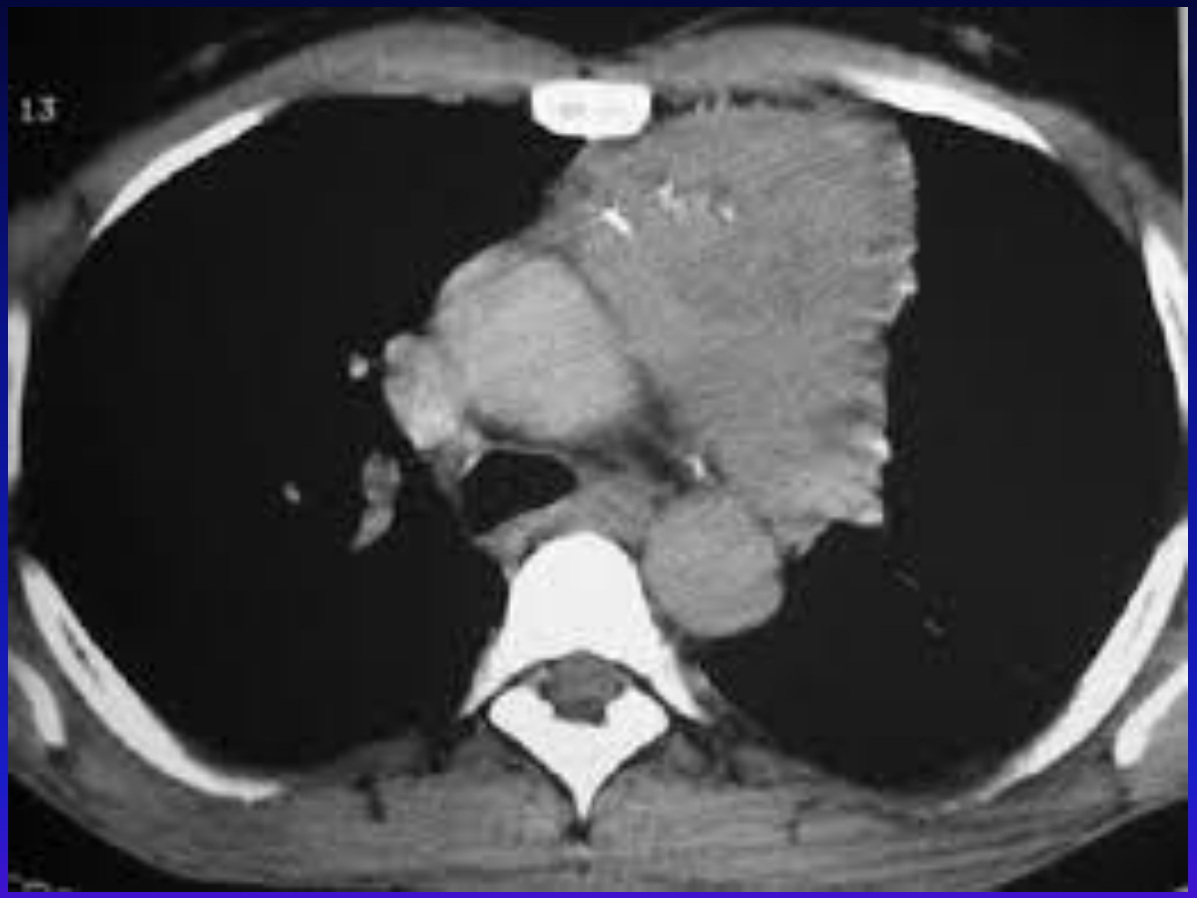

***Noninvasive Thymoma***: well encapsulated, rounded or slightly lobular shape, and usually manifest as a solid lesion with areas of hemorrhage, necrosis, or cystic degeneration.



***Invasive thymomas:*** infiltration of the adjacent structures, irregular margins, areas of low attenuation, and multifocal calcification.

# Type C Thymoma (Thymic Carcinoma)

- Has cytologic and histologic features pathognomonic of malignancy and present a strong tendency toward *early local invasion and metastatic spread.*
- Rare, predominantly in middle-aged men.
- Calcification in 10% to 40% and mediastinal LAP is seen in 40% of patients.
- Usually not associated with MG.
- Most patients are asymptomatic at diagnosis.
- The prognosis is worse and depends on stage, histology, and grading.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**Tab. 74-3. Koga Modification of the Masaoka Staging System**

| | |
|---|---|
| Stage I | Macroscopically and microscopically completely encapsulated |
| Stage IIA | Microscopic transcapsular invasion |
| Stage IIB | Macroscopic invasion into the surrounding mediastinal fat tissue or grossly adherent to, but not through, the mediastinal pleura |
| Stage III | Invasion of the neighbouring organs |
| Stage IVA | Pleural or pericardial dissemination |
| Stage IVB | Lymphogenous or haematogenous metastases |

# Prognostic Factors

- Staging and histology
- Completeness of resection
- Tumor diameter
- Paraneoplastic syndromes
  - MG symptoms , earlier diagnosis
  - Acquired hypogammaglobulinemia and pure red cell aplasia cause significant morbidity
- Involvement of great vessels (stage III)
- recurrence

# Treatment

- Surgery
- Chemotherapy
  - For recurrent, metastatic, and locally advanced diseases.
- Radiotherapy
  - No need for those stage I after complete resection

# Germ cell tumors

- Group of tumors with histology identical to that of some neoplasms of the testicle and the ovary, all of which are believed to derive from common primary germ cells.
- The mediastinum, is the most common site for extragonadal localization.
- 10% to 15% of masses of the anterior mediastinum.
- Manifest in young adults in the 3rd -5th decade.

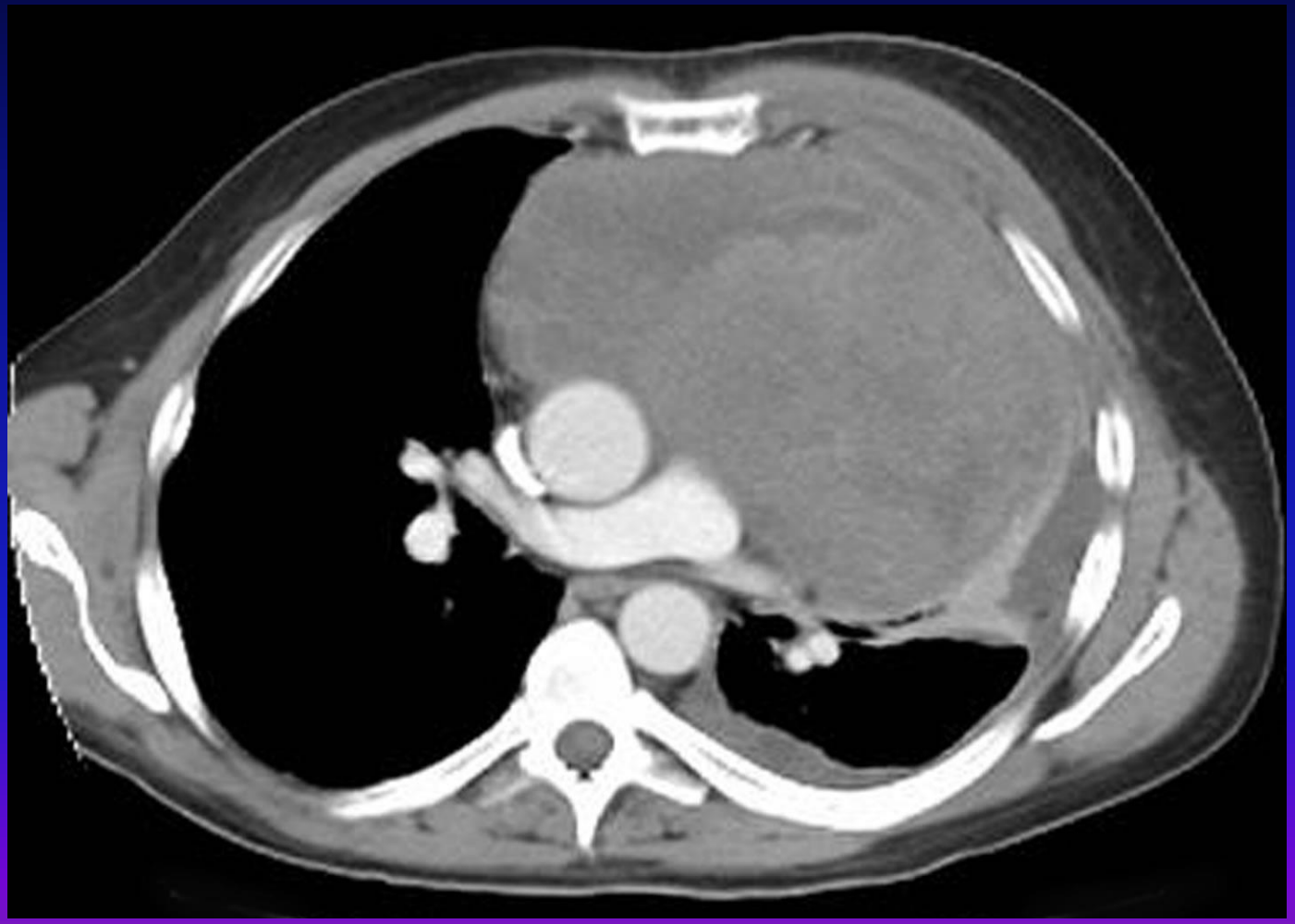
# Germ cell tumors

- Classification:
  - Benign tumors (80%) are mature teratomas,
  - Malignant include:
    - Seminomas.
    - Nonseminomatous Germ cell tumors
    - Malignant (immature) teratomas
- Associated with Klinefelter's syndrome in 8% of cases and hematologic malignancies.
- Testing of serum α-fetoprotein and (β-HCG) is mandatory

**Tab. 73-1. Classification of Mediastinal Germ Cell Tumours**

| | |
|---|---|
| Teratoma | Mature teratoma<br>Immature teratoma<br>Teratoma with malignant transformation |
| Seminoma | |
| Non-seminomatous germ cell tumours | Yolk sac tumour<br>Embryonal carcinoma<br>Choriocarcinoma<br>Mixed germ cell tumours |

# NSGST



# Teratoma

- Almost exclusively found in the anterior mediastinum.
- Often diagnosed in 3rd and 4th decade.
- ♀=♂
- May have a solid or cystic appearance.
- Sectioned surface may show hair, teeth, or seb material. Most common component is skin and its appendages.
- Mature teratomas are the most common variant, accounting for 70% of mediastinal germ cell tumors in children and 60% in adults.

# Teratoma

- They are well delimited in relation to the surrounding mediastinal structures and may be cystic.
- Incidentally discovered. However, they can reach a remarkable size and can give rise to local symptoms.
- Histologically, mature teratomas consist of irregularly arranged, well-differentiated adult tissues of ectodermal, mesodermal and endodermal origin.

# Teratoma

*Diagnosis*

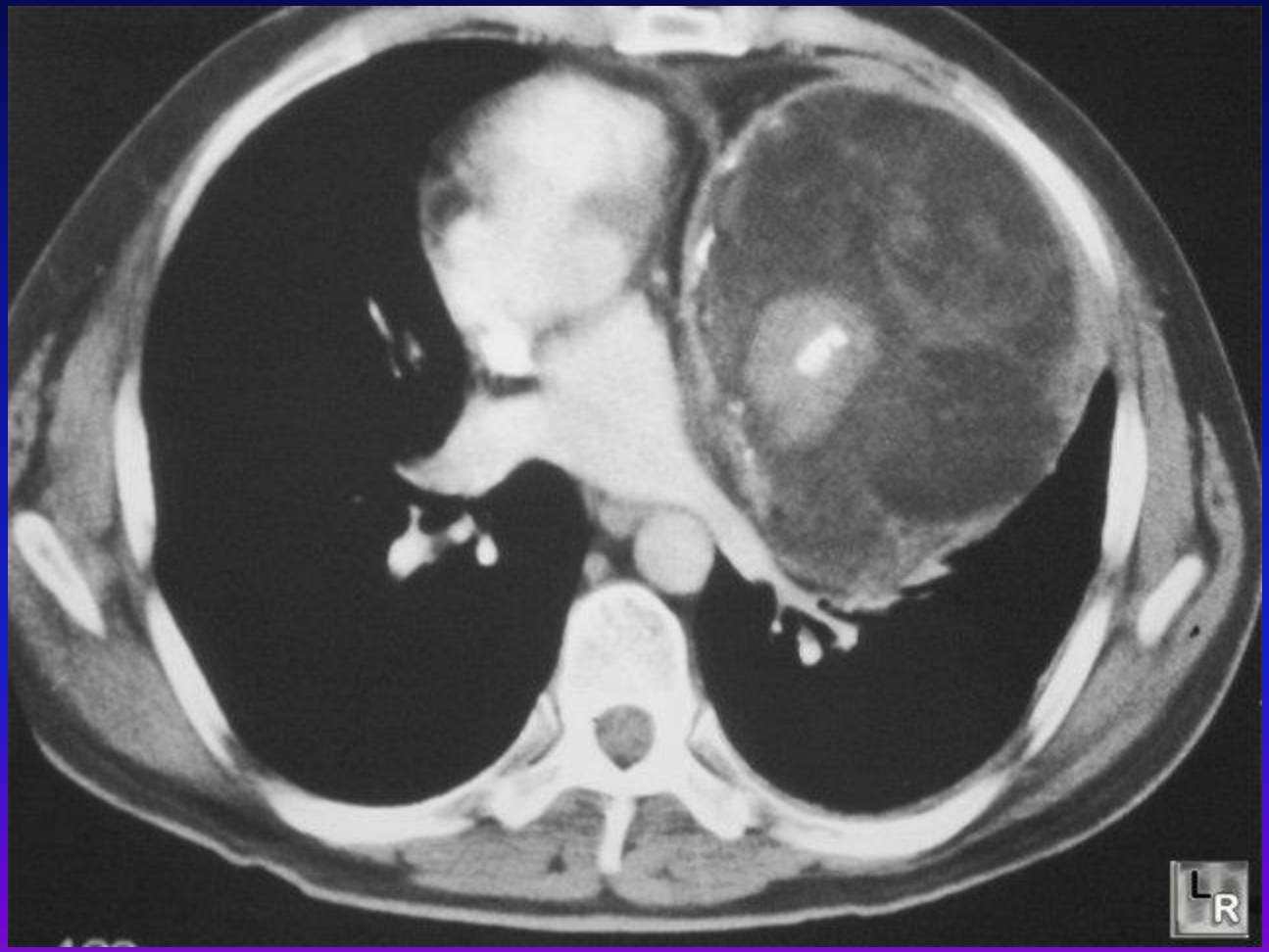
CT of the chest:

- Well-defined, smooth or lobulated margins.
- They are encapsulated and display heterogeneous attenuation due to the combination of soft tissue, fluid, fat, and calcific components.
- They are typically multilobulated cystic tumors with walls of varying thickness. Cough productive of hair or sebum is a pathognomonic sign of rupture into the tracheobronchial tree.

- ***Treatment:***

  Surgical excision

# Teratoma



# Teratoma

- Immature teratomas are made up of the same differentiated tissues as mature forms in association with poorly organized fetal-type tissue.
- In childhood, prognosis is good, whereas at any other age, their behavior is often aggressive.

# Teratoma

- Teratomas with malignant transformation, teratocarcinomas, contain a malignant component, most commonly sarcoma.
- These tumors tend to be larger than benign forms and are often found to invade adjacent structures at the time of diagnosis.

# Lymphoma

- Is a relatively common mediastinal tumor.
- 20% of all mediastinal tumors in adults and 50% in children.
- They are mostly Hodgkin's lymphomas and seldom are confined only to the mediastinum at diagnosis.
- The most common variants of non-Hodgkin's lymphomas that primarily affect the anterior mediastinum are large B-cell lymphoma and lymphoblastic lymphoma

# Middle mediastinal masses

- ***Benign***
  - Benign adenopathy
  - Cysts
  - Esophageal mass
  - Hiatal hernia
  - Cardiac and vascular structures
  - Lipomatosis
  - Cardiophrenic fat pad
  - Foramen of morgagni hernia
  - Ectopic thyroid

- ***Malignant***
  - Lymphoma
  - Metastases
  - Esophageal cancer
  - Thyroid carcinoma

# Enlarged Lymph Nodes

- Located around the tracheobronchial tree, the lower esophagus, or the aortopulmonary window.
- DDx: malignancy (e.g., lymphoma, metastatic spread from lung cancer or other malignancies) and inflammatory lymph node changes due to infection (tuberculosis, histoplasmosis) or granulomatous disease (sarcoidosis).
- CT scans may show typical patterns such as calcifications (tuberculosis) or necrosis (lymphomas).

# Bronchogenic Cysts

- Result from abnormal budding of the tracheal diverticulum between the 3rd&6th weeks of gestation.
- 5% to 10% of all mediastinal lesions.
- Usually found adjacent to the tracheobronchial tree but can be also found in the posterior mediastinum or within the lungs.
- Cause symptoms in adults in 30% to 45% of cases. Chest pain is MC presenting symptom.

# Bronchogenic Cysts

- At CT, well-defined, round masses with a homogeneous density similar to water.
- However, density and the heterogeneous aspect can make diagnosis difficult
- If there is a direct communication with the tracheobronchial tree, air-fluid levels can be seen.
- ***Treatment***

  Surgical excision

# Pericardial Cysts

- Benign intrathoracic lesions and constitute 7% of all mediastinal tumors.
- Typically located at the right cardiophrenic angle (50%-70%) or at the left cardiophrenic angle (30%-40%), or rarely in other mediastinal locations not adjacent to the diaphragm.
- Their size varies from a few centimeters to 30 cm.
- They are usually congenital but may also be acquired after cardiothoracic surgery.

# Pericardial Cysts

- Most pericardial cysts are asymptomatic.
- Patients may present with:
  - Chest discomfort
  - Dyspnea
  - Cough
  - Life-threatening complications such as cardiac tamponade have been reported.

# Pericardial Cysts

- Definitive diagnosis by use of CT may be challenging.

- Echocardiography was shown to be a superior noninvasive modality in selected cases to delineate the exact location of the cyst and to differentiate it from other potential diagnoses such as fat pad, ventricular or aortic aneurysm, and solid tumors

# Posterior mediastinal masses

***Benign***

- Neurofibroma
- Schwannoma
- Ganglioneuroma
- Pheochromocytoma
- Chemodectoma
- Foramen of bochdalek hernia
- Meningocele

***Malignant***

- Neurofibrosarcoma
- Neuroblastoma
- Ganglioneuroblastoma
- M.pheochromocytoma
- M. chemodectoma

# Neuroblastoma

- Highly malignant.
- Most common extracranial solid malignancy in pediatric patients.
- The most common intrathoracic malignancy of childhood.
- Adrenal gland is a common primary site, but 14% of all neuroblastomas arise in the thorax, where the tumors are commonly associated with extension into the spinal canal and osseous invasion.

# Neuroblastoma

- Is not as recalcitrant to chemotherapy and surgical resection as are other chest malignancies.
- They are more likely to be resectable, with less invasion of surrounding organs.
- >1/2 occur in children <2 years of age.
- 90% arise within the first decade of life

# Thank You

# Thymoma

- MG is most frequent in ♀, and symptoms include diplopia, ptosis, dysphagia, weakness, and fatigue.
- ***Diagnosis:***
  - Serum anti-acetylcholine receptor antibody test even if they are asymptomatic.
  - CT-Chest with contrast.
- ***Treatment***
  - Depends on the stage(mainly surgical resection) but radio- or radiochemotherapy have specific indications