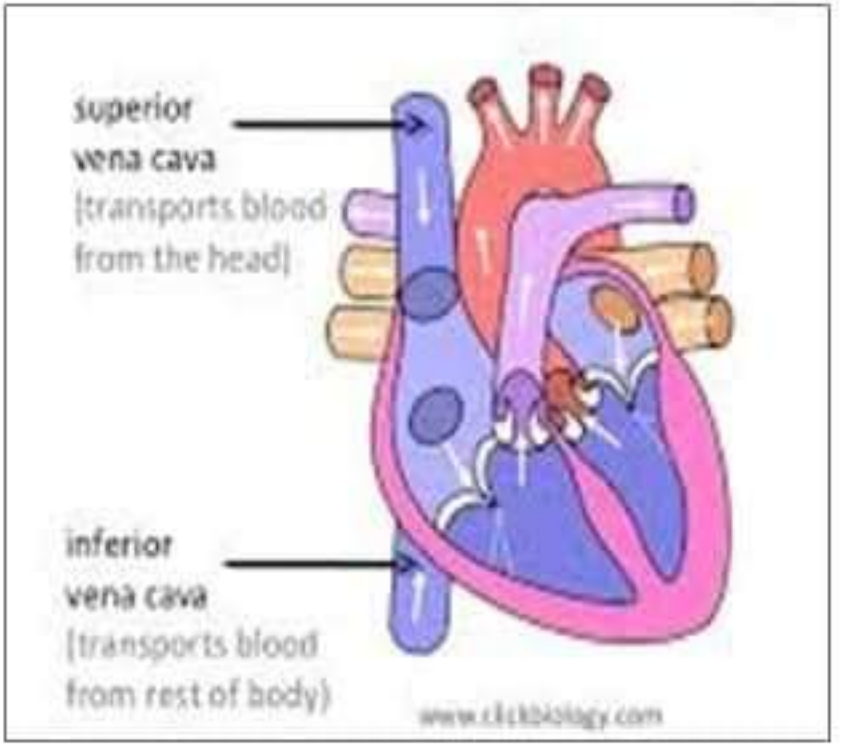
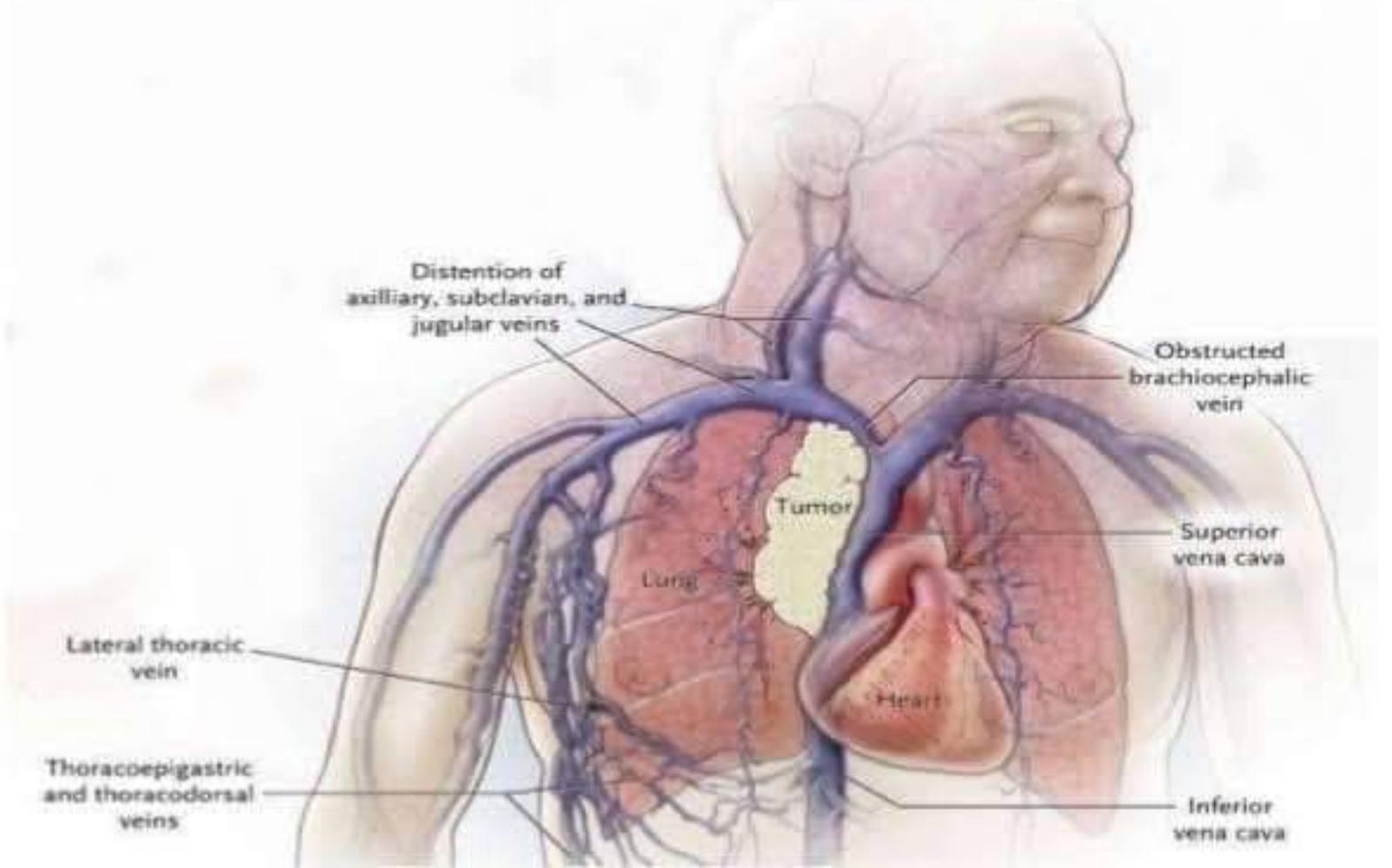
SVC syndrome

Superior vena cava

- carries venous blood from the head, arms, and upper trunk to the heart
- carries approximately one third of the venous return to the heart.



SUPERIOR VENA CAVA SYNDROME



SVC syndrome

Obstruction of Superior vena cava

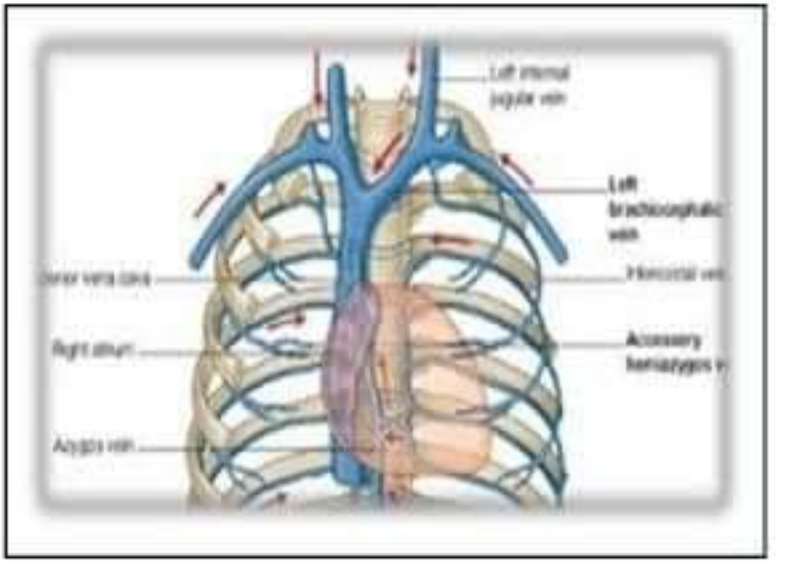
- Reduction in venous return of face, neck, upper extremities

- Collateral development of venous system
  - azygos, internal thoracic, paraspinous, esophageal

SVC syndrome

Definition

- The ***clinical manifestation*** of obstruction of the superior vena cava, with severe reduction in venous return from the head, neck, and upper extremities



SVC Obstruction

Lung Cancer – Commonest cause for SVC obstruction

- 2-5% of lung Cancer patients
  - 20% SCC develop SVC obstruction due to central airways involvement
  - Marker for poor prognosis in NSCLCA – median survival 5 months
  - Management:
    - Tissue diagnosis,
    - Chemo/Radiotherapy,
    - Endovascular stent – in refractory disease or rapid symptomatic progression

1. **Lymphoma**
   - Non Hodgkins Lymphoma
2. **Mediastinal metastasis**
   1. Germ Cell Cancer
   2. Metastastic Breast Cancer
   3. Thymoma
3. **Non Malignant Causes**
   1. Mediastinal Fibrosis – Endovascular stent, SVC bypass surgery
   2. Thrombosis of indwelling catheter devices e.g. Pacemaker

Classification of SVCS

- There are three main classification proposals which follow different methods of categorization.

Doty and Standford's classification (anatomical)

1. **Type I**: stenosis of up to 90% of the supra-azygos SVC
2. **Type II**: stenosis of more than 90% of the supra-azygos SVC
3. **Type III**: complete occlusion of SVC with azygos reverse blood flow
4. **Type IV**: complete occlusion of SVC with the involvement of the major tributaries and azygos vein

Pathophysiology

- Pathogenetic basis of SVCS is obstruction to the blood flow.
- It can result from intrinsic or extrinsic obstacles.
  - Intrinsic-thrombosis or invading tissue(uncommon).
  - Extrinsic factors- compression or stricture of the vein.
- When obstruction of the SVC occurs,
  - Vascular resistances rise and the venous return decreases.
  - SVC pressure increases consistently.
  - SVC with significant stenosis- collateral circulation opens up to bypass the obstruction and restore the venous return.

BOX 1: PRESENTING SYMPTOMS OF SVCO

Common

- Facial swelling
- Arm swelling
- Distended neck and chest veins
- Facial plethora
- Shortness of breath
- Cough
- Hoarseness
- Syncope

Less common

- Stridor
- Headaches
- Dizziness
- Visual symptoms
- Confusion
- Coma

SUPERIOR VENA CAVA SYNDROME

1. Dilated veins on neck and chest.

2. Jugular engorgement

3. Oedema and conjunctival haemorrhage.

4. Suffused (flushed) face.

5. Tongue angiomata.

Any specific physical sign?

- **Pemberton Sign**
  - Exaggeration of edema and flushing with placement of the patient's arms overhead
- Indicates compression of vascular structures in the thoracic inlet
  - Highly indicative of SVCO
  - Substernal goitre

Diagnosis

- **D**iagnosis of SVCS can be made simply on physical examination.
- When the extent of disease is minimal, the physical findings may not be prominent then it is difficult to diagnose.
- Establishing the underlying etiology is more important because certain disorders that cause SVCS may be more amenable to specific treatment regimens.

  SCLC and lymphoma -Chemotherapy/irradiation, thrombosis does not respond to this treatment.

Superior Vena Cava Syndrome

Treatment

- SVC syndrome is **not true medical emergency**.
- Biopsies should be performed before instituting therapy.
  - ✓ Mediastinoscopy, bronchoscopy, biopsy
- Stenting of the SVC
- Radiotherapy is a standard treatment modality.
- Chemotherapy and corticosteroids can also be used.

Treatment Of SVCO

- Depending on the underlying condition, multiple treatment options are available for superior vena cava obstruction. The primary treatment options include
- **Medical Care**
- Radiation
- Chemotherapy
- Thrombolytic therapy
- Anticoagulation
- Stents and balloon angioplasty and
- Surgery.

Radiation therapy

Indications.

- The majority of cases of SVCS are caused by malignancy; thus, most patients receive radiation treatment at some point in their illness.

Emergency radiation treatment

- To life-threatening cerebral or laryngeal edema prior to a tissue diagnosis of malignancy.
- To relieve obstructive symptoms
- Inappropriate for the treatment of an underlying thrombosis or granulomatosis causing the obstruction

Superior Vena Cava Syndrome (9)

- Small cell lung cancer – Chemotherapy alone or in combination with thoracic irradiation
- Non small cell lung cancer – Radiotherapy
- Non-Hodgkin lymphoma – complete relief of SVCS symptoms within 2 weeks of the onset of any type of treatment (chemotherapy, chemoradiation, radiotherapy)
- Non malignant causes – mediastinal granuloma that was attributed histoplasmosis
- Catheter-Induced Obstruction – thrombosis

Supportive care and medical management

- No data documenting the effectiveness of this maneuver, the head should be raised to decrease hydrostatic pressure and head and neck edema.
- Obstruction of blood flow through the SVC slows venous return. This can result in local irritation or thrombosis of veins in the upper extremities, or delayed absorption of drugs from the surrounding tissues. Thus, the **use of intramuscular injections in the arms should be avoided**.
- For patients who have obstruction of the SVC resulting from intravascular thrombus associated with an indwelling catheter, **removal of the catheter is indicated**, in conjunction with systemic anticoagulation.