GOLGI COMPLEX

- The Golgi apparatus or the Golgi body or Golgi complex or simply Golgi is a cellular organelle present in most of the cells of the eukaryotic organisms.
- It is referred to as the manufacturing and the shipping center of the cell.
- Golgi is involved in the packaging of the protein molecules before they are sent to their destination. These organelles help in processing and packaging the macromolecules like **proteins** and lipids that are synthesized by the cell and hence act as the 'post office' of the cell.
- Golgi apparatus was discovered in the year 1898 by an Italian biologist Camillo Golgi.

Structure of Golgi Apparatus

- Under the electron microscope, the Golgi apparatus is seen to be composed of stacks of flattened structures that contain numerous vesicles containing secretory granules.
- The Golgi apparatus is morphologically very similar in both plant and animal cells. However, it is extremely pleomorphic in some cell types.
- Typically, however, Golgi apparatus appears as a complex array of interconnecting tubules, vesicles and cisternae.

The golgi is made of 5-8 folds called **cisternae**. The cisternae contain specific enzymes creating five functional regions which modify proteins passing through them in a stereotypical way, as follows:

- **Cis-Golgi network**: faces the nucleus, forms a connection with the endoplasmic reticulum and is the entry point into the Golgi apparatus.
- **Cis-Golgi**: major processing area allowing biochemical modifications
- **Medial-Golgi**: major processing area allowing biochemical modifications
- **Trans-Golgi**: major processing area allowing biochemical modifications
- **Trans-Golgi network**: exit point for vesicles budding off the Golgi surface, packages and sorts biochemicals into the vesicles according to their destination.

- ***Cisternae:*** It is simplest unit of the Golgi apparatus.

- Cisternae are central, flattened, plate like or saucer shaped closed compartments that are held in parallel bundles or stacks one above the other.
- In each stack, cisternae are separated by a space of 20-30 nm which may contain rod like structures or such fibres.
- Each stack of cisternae forms a dictyosome which may contain 5-6 Golgi cisternae in animal cells or 20 or more in plant cells.

- Each cistern is bounded by a smooth unit membrane (7.5 nm thick), having alumen varying in width from about 500 to 1000 nm.
- The margins of each cistern are generally curved so that the entire Dictyosome of the Golgi apparatus takes on a bow-like appearance.
- The cisternae at the convex end of the Dictyosome comprise proximal, forming or cis - face and the cisternae at the concave end of the Dictyosome form the distal or trans- face.

➢ ***Tubules:***

A complex array of associated vesicles and anastomosing tubules (30-50 nm) surrounded the Dictyosome and radiate from it. In fact, the peripheral area of the Dictyosome is fenestrated in structure.

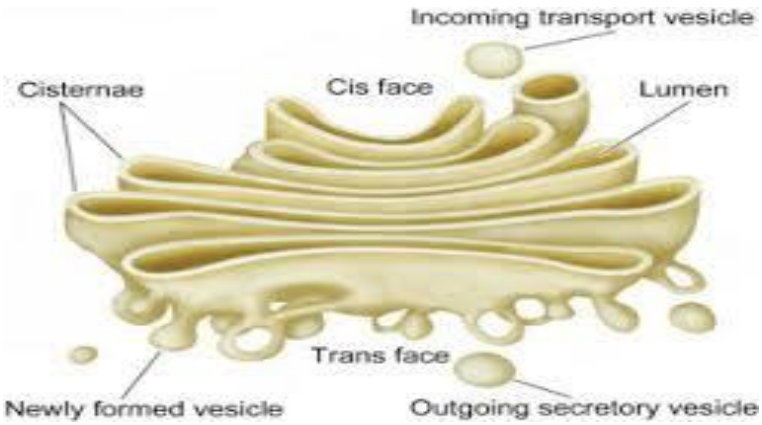
➢ ***Vesicles:***

The vesicles are of 3 types and these are almost 60 nm in diameter.

a) Transitional vesicles: these are small membrane bound vesicles which are thought to form as blebs from the transitional ER to migrate and converge to cis face of Golgi, where they coalesce to produce new cisternae.

b) Secretory vesicles: these are varied sized membrane bound vesicles that discharge from margins of cisternae of Golgi. They, often, occur between the maturing face of Golgi and the plasmalemma.

c) Clathrin-coated vesicles: these are spherical structures about 50 micrometers in diameter and with a rough outer surface. They are found at the periphery of the organelle, usually at the ends of single tubules and are morphologically quite distinct from the secretory vesicles. The clathrin-coated vesicles are known to play an important role in the GERL system.



Functions of Golgi apparatus:

The Golgi apparatus has the following important roles-

a) **Biosynthetic activities:** As for example, it contains enzymes that complete the glycosylation of glycoproteins and glycolipids. Again, proteases complete the hydrolytic events that convert inactive proteins to their active conformations. For example, insulin is produced and secreted by beta cells of the islets of Langerhans in the pancreas and actually it becomes activated from proinsulin by the help of Golgi apparatus in those cells.

b) **Transfer of proteins to the ER:** Proteins and enzymes that function in the ER but are transferred to the Golgi body are returned to it from the CGN. Receptors in the CGN recognize the ER retrieval signals Lys-Lys-X-X and KDEL on membrane and soluble proteins respectively. The proteins are concentrated into COP I coated vesicles by the CGN for return to the ER. The CGN then release these vesicles to return the proteins to the ER.

c) **Secretion of materials from the cell:** The TGN packages materials for secretion from the cell into appropriate secretory vesicles. These materials include proteolytic enzymes and hormones from a variety of secretory cells. The Golgi apparatus also has an essential role in delivering enzymes to the endosomal-lysosomal system.

d) **Distribution of integral membrane proteins:** Integral membrane proteins are synthesized by ribosomes of the RER and are inserted into the membrane of the RER after synthesis. Then these proteins re transported to the Golgi apparatus. Then the Golgi body sorts the proteins into appropriate COP I and clathrin coated vesicles, which transport the membrane proteins to their destinations. Fusion of the vesicles with the target membranes integrates the protein into that membrane.

Clinical Relevance - Wilson's Disease

Wilson's disease **is an autosomal recessive disorder characterised by an abnormal accumulation of copper in the body. The liver is particularly susceptible to this accumulation.**

One of new emerging Golgi functions consists in the regulation of copper homeostasis by coordinating the relocation and activity of copper transporters. Of these, the Cu-transporting ATPase ATP7B (known as Wilson disease protein) plays a key role in the maintenance of the Cu balance in the body *via* the supply of essential Cu to the systemic circulation and *via* elimination of excess Cu into the bile. Mutation of the ATP7B gene is implicated in the development of Wilson disease. The ATP7B gene codes for copper-transporting ATPase 2 (also known as Wilson disease protein or WDP), an enzyme located in the Golgi apparatus of many cells such as **hepatocytes** and **neurones**.

Patients experience symptoms and signs of **liver failure**, such as itching, abdominal swelling, fatigue, jaundice, ascites and Kayser-Fleischer rings (deposition of copper in the cornea).

They may also present with **neuropsychiatric** symptoms such as confusion, psychosis, personality changes and tremor. Copper may also accumulate in joints causing joint pain and swelling. Diagnosis is based on abnormal liver function tests, **serum copper** levels, urinary copper levels, and serum caeruloplasmin levels. If the diagnosis is unclear a liver biopsy may be indicated.

Management is aimed at reducing copper levels using copper chelating agents such as **penicillamine**.

References:

- **Verma and Agarwal, 2006.**
- **Alberts, 2004.**
- **Wang et al., 2013**
- **Polishchuk, 2019.**