# The microstructure of a cell

The microstructure of a cell is made up of a complex system of organelles. Each organelle has an important function with regards to the production and transport of information into the cystic fibrosis transmembrane conductance Regulator (CFTR) protein. The nucleus contains DNA, the coded information required for protein synthesis. The genetic information coded within the nucleus is transcribed and exported via mRNA to associate with the ribosomes [1] . The nucleus also controls the formation of ribosomes. Ribosomes are made up of DNA and protein and are found throughout the cytoplasm of a cell, either free or bound to the endoplasmic reticulum (ER). The ribosome is the site of CFTR protein synthesis, which occurs when the mRNA is translated into a sequence of amino acids and ultimately a protein is synthesised.

The ER is a complex system of interconnected tubules and vesicles and consists of rough endoplasmic reticulum (RER) and smooth endoplasmic reticulum (SER). The RER is the centre for protein synthesis (translation) as it is on the RER that ribosome’s can be found. The RER is important for the transportation of proteins via vesicles to the Golgi complex [2] .

The SER has no bound ribosomes on its surface. It is made up of a branched tubular network which increases its surface area. This large surface area is suited for action and storage of enzymes and enzyme products.

The Golgi complex is arranged into cisternae, which are membrane bound sacs. It has a close relationship with the ER and is designed to act as an export site. The Golgi complex is responsible for the processing and transport in vesicles of cell materials, enzymes and proteins. It is also

responsible for the movement of proteins from cis to trans face. The Golgi is also involved in the control of secretions and lysosome formation.

The lysosomes function as degradation vesicles, degrading bacteria, unwanted organelles, such as mitochondria, and other unwanted cellular products. They contain hydrolases with degrade nucleotides, proteins and lipids [3] .

Peroxisomes are also degradation vesicles that are evident in the cytoplasm although these are not made in the Golgi complex. There key function is to remove hydrogen peroxide from the body. Peroxisomes also contain enzymes important for protein synthesis.

The most significant organelle is the mitochondria, which is the site of aerobic respiration in a cell and the production of ATP. The energy produced is transferred to ATP and this energy is then available for all cellular functions. It is therefore evident that each organelle has a significant function in the various stages of the production of the CFTR protein.

The CFTR protein spans the membrane of a cell and acts as an ion channel. Globular integral proteins than span the membrane of the cell allow the passage of the CFTR protein into the cell [4] . The membrane of a cell is referred to as the fluid mosaic model. It has a lipid bilayer which forms the structure of the membrane and is responsible for the fluidity of the membrane. The fluid mosaic model structure also contains phospholipids, proteins and glycoproteins. The surface of the membrane is formed by water soluble (hydrophilic) heads. The interior of the membrane is formed by water

insoluble (hydrophobic) tails. Lipid soluble molecules can pass through the membrane but water soluble molecules are unable to pass through. Globular integral proteins that span the membrane allow passage of water soluble molecules and ions into and out of the cell [5] . Other proteins act as carrier proteins to transfer substances across the cell membrane that would be unable to cross alone.

The CFTR protein spans the membrane of the cell and acts as a chloride ($Cl^-$) ion channel. The CFTR also regulates sodium ($Na^+$) ion transport. The globular integral proteins spanning the membrane allow the CFTR protein to pass through, transporting $Cl^-$ ions across the membrane. The CFTR protein is important in maintaining salt and water balance in the cells of the airways and lungs. As the $Cl^-$ ion concentration in the epithelial cells increases a concentration gradient is created. The $Cl^-$ ions then diffuse via osmosis out of the cell via the CFTR protein into the mucus. At the same time $Na^+$ ions are drawn out of the cell to maintain equilibrium. The combined effects of $Cl^-$ ions and $Na^+$ ions allow water to diffuse out of the cell which then dilutes the mucus on the surface of the cell membranes.

Individuals with cystic fibrosis have a blocked or absent CFRT protein. The CFTR protein becomes stuck in the ER and Golgi complex which are important organelles for the synthesis and transport of the protein to the cell membrane. Therefore the CFTR is not processed effectively; it is not transported to the cell membrane and therefore cannot perform adequately within the cell. This causes the cell membrane to become impermeable to $Cl^-$ ions [6] . The $Cl^-$ ions build up within the cell as they are unable to move out

of the cells via the CFTR protein, therefore the ion concentration within the cell increases. The $Na^+$ ion transport within the cell is also affected. As the ion concentration increases, water is drawn out of the surface liquid and mucus membrane into the cell via osmosis therefore causing the mucus to become thick and sticky. The vital function of the CFTR protein to control the regulation of water in the cells and to maintain the fluidity of the mucus is then defective.

The respiratory tract is lined with mucus which acts as a protective barrier against pathogens and irritants. Antibodies are also secreted into the mucus which destroys any pathogens. Cilia also have an important role to play in maintaining a healthy respiratory system. Cilia are tiny projections which line the respiratory tract. They beat rapidly to sweep the mucus and any pathogens or irritants along the respiratory tract towards the throat where it is swallowed and destroyed by enzymes in the stomach.

Individuals with cystic fibrosis are prone to respiratory infections due to the viscous consistency of the mucus lining the airways and lungs. The thick mucus also makes it difficult to get air in and out of the lungs as the airways become clogged. The mucus is too thick to function normally and the antibodies are unable to destroy the pathogens effectively. The cilia are unable to move the thick mucus along the respiratory tract, therefore pathogens and irritants are trapped within the mucus. This leads to chronic and repeated chest infections and breathing difficulties. Persistent coughing to remove the irritants can also lead to pneumothorax and bleeding in the

lungs. Eventually the lung tissue becomes fibrosis [7] , elasticity is lost causing difficulties with lung inflation and extreme breathing difficulties.

Bibliography.

Alberts B, Bray D, Hopkin K, Johnson A, Lewis J, Raff M, Roberts K & Walter P(2004) Essential Cell Biology. Garland Science, New York.

Elliot, W H & Elliot, D C (2004) Biochemistry & Molecular Biology. Oxford University Press, Oxford

Seeley R, Stephens, TD & Tate P (2000) Anatomy & Physiology (5 th Edition), McGraw Hill, USA

Sherwood L, (1996) Fundamental of Physiology A human Perspective (2 nd Edition), West, USA

---

Footnotes

[1] Alberts B, Bray D, Hopkin K, Johnson A, Lewis J, Raff M, Roberts K, Walter P (2004) chpt. 2

[2] Elliot, W & Elliot, C D (2004)

[3] Sherwood, L (2000) p. 21

[4] Elliot (ibid)

[5] Seeley, Stephens & Tate (1996) p. 60

[6] Sherwood (ibid) p. 41

[7] Sherwood (ibid) p. 41