MICRO CONNECTIONS - Primary Structure, Dysfunctional Proteins, and Cystic Fibrosis

Proteins associated with the plasma membranes of cells are classified as peripheral or integral. Peripheral proteins are associated with one side of the membrane, whereas integral proteins are embedded in the membrane. Integral proteins can allow specific materials to move into or out of the cell. Cystic fibrosis (CF) is a human genetic disorder caused by a change in an integral membrane protein. It affects mostly the lungs but may also affect the pancreas, liver, kidneys, and intestine. Individuals who have CF are unable to make a transmembrane (integral) protein (CFTR) that usually helps transport salt and water into and out of cells (Figure 3.34). Because of a mutation in the DNA, one amino acid, phenylalanine, is left out when the integral transport protein is made. The loss of one amino acid changes the primary structure of the protein.

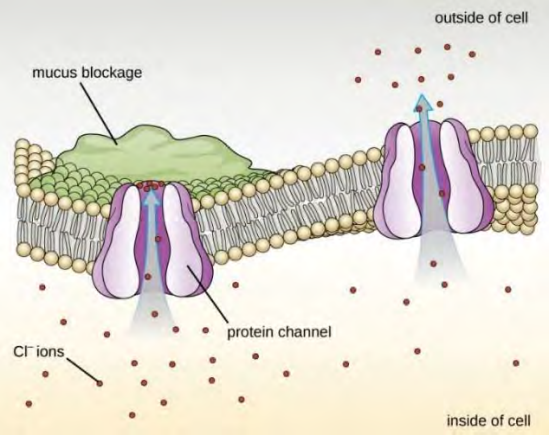

Figure 3.34 The normal CFTR protein is a channel protein that helps salt (sodium chloride) move in and out of cells. (credit: Parker et al. / Microbiology OpenStax)

The change in the primary structure prevents the protein from functioning correctly, which causes the body to produce unusually thick mucus that clogs the lungs and leads to the accumulation of sticky mucus. The mucus obstructs the pancreas and stops natural enzymes from helping the body break down food and absorb vital nutrients.

In the lungs, the altered mucus provides an environment where bacteria can thrive. This colonization leads to the formation of biofilms in the small airways of the lungs. The most common pathogens found in the lungs of patients with cystic fibrosis are *Pseudomonas aeruginosa* (Figure 3.35) and *Burkholderia cepacia*. *Pseudomonas* differentiates within the biofilm in the lung and forms large colonies, called "mucoid" *Pseudomonas*. The colonies have a unique pigmentation that shows up in laboratory tests (Figure 3.35) and provides physicians with the first clue that the patient has CF. Such colonies are rare in healthy individuals.

Figure 3.35 (a) A scanning electron micrograph shows the opportunistic bacterium Pseudomonas aeruginosa. (b) Pigment-producing P. aeruginosa on cetrimide agar shows the green pigment called pyocyanin. (credit a: modification of work by the Centers for Disease Control and Prevention / Microbiology OpenStax)

CONCEPTS IN ACTION - For more information about cystic fibrosis, visit the Cystic Fibrosis Foundation website.

Section Summary

Proteins are a class of macromolecules that can perform a diverse range of functions for the cell. They help in metabolism, provide structural support, speed up the rate of chemical reactions, transport materials, and function as hormones. The building blocks of proteins are amino acids. Proteins have four structures: primary, secondary, tertiary, and quaternary. Protein shape and function are intricately linked. Any change in shape caused by changes in temperature, pH, salinity, or chemical exposure may lead to protein denaturation and a loss of function.

Exercises

1. A _____ bond forms between the carboxyl group of one amino acid and the amino group of another amino acid.
    a. hydrogen
    b. ionic
    c. peptide
    d. all of the above

2. Enzymes speed up chemical reactions by _____ the energy needed to start the reaction.
    a. increasing
    b. decreasing

3. Denaturation can sometimes be reversed.
    a. True
    b. False

4. The monomers that make up proteins are called _____.
    a. nucleotides
    b. disaccharides
    c. amino acids
    d. chaperones

5. Explain what happens if even one amino acid is substituted for another in a polypeptide chain.
6. A mysterious disease results in the unfolding of proteins. This disease effects which protein structure?

Answers

1. (c)
2. (b)
3. (a)
4. (c)
5. This causes a change in protein structure and function.
6. Tertiary structure is affected when proteins unfold. This will also affect the quaternary structure.

Glossary

amino acid: a monomer of a protein

denaturation: loss of shape in a protein that may be a result of changes in temperature, pH, or chemical exposure

enzyme: a catalyst in a biochemical reaction that is usually a complex or conjugated protein

hormone: a chemical signaling molecule, usually a protein or steroid, secreted by an endocrine gland or group of endocrine cells; acts to control or regulate specific physiological processes

hydrophilic: describes a substance that dissolves in water; water-loving

hydrophobic: describes a substance that does not dissolve in water; water-fearing

peptide bond: a covalent bond that forms between one amino acids carboxyl group and another amino acids amino group

polypeptide chain: a long chain of amino acids linked by peptide bonds

primary structure: a linear sequence of amino acids in a protein

protein: a biological macromolecule composed of one or more chains of amino acids

quaternary structure: association of different polypeptide chains in a protein

secondary structure: structure that proteins form by hydrogen bonding between the oxygen atom of one amino acid and the hydrogen attached to the nitrogen atom of another amino acid

substrate: a reactant that binds to a specific enzyme

tertiary structure: a protein's three-dimensional conformation, including interactions between secondary structural elements