

GLUCOSE-6-PHOSPHATE DEHYDROGENASE DEFICIENCY IN THE BODY

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Annotation: Glucose-6-phosphate dehydrogenase deficiency, hereditary metabolic defect characterized by an increased tendency of the red blood cells to break and release their hemoglobin (hemolysis), especially after the intake of certain drugs. The condition is caused, as the name indicates, by the markedly reduced activity in the red blood cells of a particular organic catalyst, or enzyme, called glucose-6-phosphate dehydrogenase.

Key words: Glucose-6-phosphate, enzyme, protein, hemoglobin.

This low enzyme activity is associated with a decrease in the formation of certain substances that normally help to prevent the oxidative destruction of the red blood cell membrane. Under normal conditions, the affected red blood cells are only slightly more fragile than usual, but more than 40 drugs, including chloramphenicol and sulfonamides, all of which are converted in the body to oxidant compounds, have been shown to produce hemolysis in susceptible persons. There seem to be several variants of the disorder, all of which appear to be sex-linked and fully expressed in males only. The most common form is found chiefly in persons whose ancestors inhabited either Africa or the Eastern Mediterranean basin. A possible protective effect of this metabolic abnormality against malaria has been suggested.

Hemoglobin, iron-containing protein in the blood of many animals—in the red blood cells (erythrocytes) of vertebrates—that transports oxygen to the tissues. Hemoglobin forms an unstable reversible bond with oxygen. In the oxygenated state, it is called oxyhemoglobin and is bright red; in the reduced state, it is purplish blue.

Hemoglobin develops in cells in the bone marrow that become red blood cells. When red cells die, hemoglobin is broken up: iron is salvaged, transported to the bone marrow by proteins called transferrins, and used again in the production of new red blood cells; the remainder of the hemoglobin forms the basis of bilirubin, a chemical that is excreted into the bile and gives the feces their characteristic yellow-brown colour.

Each hemoglobin molecule is made up of four heme groups surrounding a globin group, forming a tetrahedral structure. Heme, which accounts for only 4 percent of the weight of the molecule, is composed of a ringlike organic compound known as a porphyrin to which an iron atom is attached. It is the iron atom that binds oxygen as the blood travels between the lungs and the tissues. There are four iron atoms in each molecule of hemoglobin, which accordingly can bind four molecules of oxygen. Globin consists of two linked pairs of polypeptide chains. Hemoglobin S is a variant form of hemoglobin that is present in persons who have sickle cell anemia, a severe hereditary form of anemia in which the cells become crescent-shaped when oxygen is lacking. The abnormal sickle-shaped cells die prematurely and may become lodged in small blood vessels, potentially obstructing the microcirculation and leading to tissue damage. The sickling trait is found mainly in people of African descent, though the disease also occurs in persons of Middle Eastern, Mediterranean, or Indian descent.

Enzyme, a substance that acts as a catalyst in living organisms, regulating the rate at which chemical reactions proceed without itself being altered in the process. A brief treatment of enzymes follows. For full treatment, *see* protein: Enzymes.

The biological processes that occur within all living organisms are chemical reactions, and most are regulated by enzymes. Without enzymes, many of these reactions would not take place at a perceptible rate. Enzymes catalyze all aspects of cell metabolism. This includes the digestion of food, in which large nutrient molecules (such as proteins, carbohydrates, and fats) are broken down into smaller molecules; the conservation and transformation of chemical energy; and the construction of cellular macromolecules from smaller precursors. Many inherited human diseases, such as albinism and phenylketonuria, result from a deficiency of a particular enzyme.

Enzymes also have valuable industrial and medical applications. The fermenting of wine, leavening of bread, curdling of cheese, and brewing of beer have been practiced from earliest times, but not until the 19th century were these reactions understood to be the result of the catalytic activity of enzymes. Since then, enzymes have assumed an increasing importance in industrial processes that involve organic chemical reactions. The uses of enzymes in medicine include killing disease-causing microorganisms, promoting wound healing, and diagnosing certain diseases. All enzymes were once thought to be proteins, but since the 1980s the catalytic ability of certain nucleic acids, called ribozymes (or catalytic RNAs), has been demonstrated, refuting this axiom. Because so little is yet known about the enzymatic functioning of RNA, this discussion will focus primarily on protein enzymes.

A large protein enzyme molecule is composed of one or more amino acid chains called polypeptide chains. The amino acid sequence determines the characteristic folding patterns of the protein's structure, which is essential to enzyme specificity. If the enzyme is subjected to changes, such as fluctuations in temperature or pH, the protein structure may lose its integrity (denature) and its enzymatic ability. Denaturation is sometimes, but not always, reversible.

Bound to some enzymes is an additional chemical component called a cofactor, which is a direct participant in the catalytic event and thus is required for enzymatic activity. A cofactor may be either a coenzyme—an organic molecule, such as a vitamin—or an inorganic metal ion; some enzymes require both. A cofactor may be either tightly or loosely bound to the enzyme. If tightly connected, the cofactor is referred to as a prosthetic group. An enzyme will interact with only one type of substance or group of substances, called the substrate, to catalyze a certain kind of reaction. Because of this specificity, enzymes often have been named by adding the suffix “-ase” to the substrate's name (as in urease, which catalyzes the breakdown of urea). Not all enzymes have been named in this manner, however, and to ease the confusion surrounding enzyme nomenclature, a classification system has been developed based on the type of reaction the enzyme catalyzes. There are six principal categories and their reactions:

- (1) oxidoreductases, which are involved in electron transfer;
- (2) transferases, which transfer a chemical group from one substance to another;
- (3) hydrolases, which cleave the substrate by uptake of a water molecule (hydrolysis);
- (4) lyases, which form double bonds by adding or removing a chemical group;

- (5) isomerases, which transfer a group within a molecule to form an isomer; and
- (6) ligases, or synthetases, which couple the formation of various chemical bonds to the breakdown of a pyrophosphate bond in adenosine triphosphate or a similar nucleotide.

In most chemical reactions, an energy barrier exists that must be overcome for the reaction to occur. This barrier prevents complex molecules such as proteins and nucleic acids from spontaneously degrading, and so is necessary for the preservation of life. When metabolic changes are required in a cell, however, certain of these complex molecules must be broken down, and this energy barrier must be surmounted. Heat could provide the additional needed energy (called activation energy), but the rise in temperature would kill the cell. The alternative is to lower the activation energy level through the use of a catalyst. This is the role that enzymes play. They react with the substrate to form an intermediate complex—a “transition state”—that requires less energy for the reaction to proceed. The unstable intermediate compound quickly breaks down to form reaction products, and the unchanged enzyme is free to react with other substrate molecules. Only a certain region of the enzyme, called the active site, binds to the substrate. The active site is a groove or pocket formed by the folding pattern of the protein. This three-dimensional structure, together with the chemical and electrical properties of the amino acids and cofactors within the active site, permits only a particular substrate to bind to the site, thus determining the enzyme’s specificity.

Enzyme synthesis and activity also are influenced by genetic control and distribution in a cell. Some enzymes are not produced by certain cells, and others are formed only when required. Enzymes are not always found uniformly within a cell; often they are compartmentalized in the nucleus, on the cell membrane, or in subcellular structures. The rates of enzyme synthesis and activity are further influenced by hormones, neurosecretions, and other chemicals that affect the cell’s internal environment.

Factors affecting enzyme activity

Because enzymes are not consumed in the reactions they catalyze and can be used over and over again, only a very small quantity of an enzyme is needed to catalyze a reaction. A typical enzyme molecule can convert 1,000 substrate molecules per second. The rate of an enzymatic reaction increases with increased substrate concentration, reaching maximum velocity when all active sites of the enzyme molecules are engaged. The enzyme is then said to be saturated, the rate of the reaction being determined by the speed at which the active sites can convert substrate to product.

Enzyme activity can be inhibited in various ways. Competitive inhibition occurs when molecules very similar to the substrate molecules bind to the active site and prevent binding of the actual substrate. Penicillin, for example, is a competitive inhibitor that blocks the active site of an enzyme that many bacteria use to construct their cell walls.

Noncompetitive inhibition occurs when an inhibitor binds to the enzyme at a location other than the active site. In some cases of noncompetitive inhibition, the inhibitor is thought to bind to the enzyme in such a way as to physically block the normal active site. In other instances, the binding of the inhibitor is believed to change the shape of the enzyme molecule, thereby deforming its active site and preventing it from reacting with its substrate. This latter type of noncompetitive inhibition is called allosteric inhibition; the place where the inhibitor binds to the

enzyme is called the allosteric site. Frequently, an end-product of a metabolic pathway serves as an allosteric inhibitor on an earlier enzyme of the pathway. This inhibition of an enzyme by a product of its pathway is a form of negative feedback.

Allosteric control can involve stimulation of enzyme action as well as inhibition. An activator molecule can be bound to an allosteric site and induce a reaction at the active site by changing its shape to fit a substrate that could not induce the change by itself. Common activators include hormones and the products of earlier enzymatic reactions. Allosteric stimulation and inhibition allow production of energy and materials by the cell when they are needed and inhibit production when the supply is adequate.

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