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Review Article

Metabolic Pathology in Early Postnatal Ontogenesis

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Abstract

Metabolism in the pediatric body, especially in the period of newborns, differs significantly from the metabolism of adults. First of all, the infant is distinguished by a high energy demand due to the large specific surface area of the body, growth processes (increase in body weight) and intense muscle activity. The period of newborn ™ is characterized by flexor hypertonia, providing a high level of heat production. When screaming, crying energy consumption increases by 100-200% . A large amount of energy is spent by newborns to adapt to the conditions of extrauterine existence. Insufficient development of nervous and hormonal regulatory mechanisms, small deposition of energy materials make the regulation of metabolic processes in small children unstable, and insignificant in strength pathogenic factors easily lead to metabolic disorders.

Keywords: metabolism; pediatric body; newborns

Introduction

Metabolism in the pediatric body, especially in the period of newborns, differs significantly from the metabolism of adults. First of all, the infant is distinguished by a high energy demand due to the large specific surface area of the body, growth processes (increase in body weight) and intense muscle activity. The period of newborn ™ is characterized by flexor hypertonia, providing a high level of heat production. When screaming, crying energy consumption increases by 100-200% . A large amount of energy is spent by newborns to adapt to the conditions of extrauterine existence. Insufficient development of nervous and hormonal regulatory mechanisms, small deposition of energy materials make the regulation of metabolic processes in small children unstable, and insignificant in strength pathogenic factors easily lead to metabolic disorders.[7] Determination of the basic metabolism in a newborn child is very difficult, because it is very difficult to ensure immobility - to turn off muscle activity in a child of early age. In this regard, it is difficult to establish the optimal temperature zone, as well as to observe the nutritional conditions necessary to exclude the activity of the digestive organs and specific-dynamic action of food. Each age period has its own characteristic defined by a specific level of functioning, working capacity of the organism, metabolic rate, basic metabolism, and oxygen consumption. there is an opinion that law of “body surface”, which established the constancy of energy expenditure per unit of body surface (1 m2), a small child with a relatively large body surface expends more energy than an adult, law has exceptions; in particular, at the same size of the surface, the degree of muscle development determines the energy expenditure.[12] It is believed that in the early age period, due to the existence of flexor hypertonia of muscles, we can not talk about the basic metabolism, as all calories of food at this age go to heat formation (maintenance of homoeo-thermia) and anabolic processes. The smaller the child's body, the more water and fat it contains and the poorer the nitrogenous substances, which is associated with poor muscle development. On the first day after birth, the basic metabolism is low at 46 kcal/kg/24 h, but from the second day onward, the metabolic processes increase and increase up to 1.5 years. When calculated per unit of surface (1 m2) in a mature newborn child basic metabolism is equal to 612, in a year-old - 1100, in under-age - 400 kcal/24 h. If the incoming food does not cover the energy expenditure, starvation occurs. In the early age period, in connection with the process of intensive growth, the deficit of nutritional substances is manifested especially quickly. Caloric deficiency (incomplete starvation) in infants is found in hypolactia and in all cases

when the act of sucking is violated. Much more often we have to face a combination of incomplete and partial starvation. For the processes of growth the content of protein in food is important and at its insufficiency (protein starvation) there is a lag in the development of the child and growth retardation. [11].

Protein Metabolism

The child, except for the first 2-3 days of life, has a positive nitrogen balance. The need for infant protein per 1 kg of body weight about 3 times more than in an adult. Increase in the amount of cellular proteins - the main factor of growth. In a one-month-old child protein consumption is 2.5 g/kg, and in a 6-month-old - 4.5 g/kg. In children of the first 2 weeks of life there is maximum nitrogen retention in the body - 78.3% of nitrogenous substances of food; in a 5-month-old child, nitrogen retention is only 23.1% .Effects such as, for example, infection, trauma, pain, cold, overheating, anxiety, cause a significant and sometimes prolonged increase in the excretion of nitrogen in the urine, increasing the protein requirement by an average of 10%.Large losses of nitrogen occur in gastrointestinal disorders, fever, trauma, proteinuria, diseases with exudation, such as eczema, liver disease. Protein deficiency can occur with insufficient content in maternal milk. It is believed that, the composition of milk in women varies widely: for example, the co-fat content ranges from 1 to 8% (average 3.5-4%), and protein - from 0.7 to 1.7% (average 1.2-1.5%). Protein-calorie deficiency may occur with incomplete complementary foods. In economically underdeveloped countries as a complementary food used vegetable protein, incompletely valuable in the composition of amino acids and poorly digested [3]. The predominance of carbohydrates in food leads to water and salt retention in the skin, which creates conditions for the development of exudative diathesis, eczema, manifestations of allergies. Excessive carbohydrate diet with insufficient content of animal proteins in the food and as a consequence of the latter - hypoproteinemia - lead to water retention in the skin. This is largely promoted by the structural organization of the child's skin, a feature of which is an abundance of vessels and a large number of mesenchymal elements. Significantly less thickness of all layers of the dermis, insufficiency of pigment-forming structures, incomplete development of sweat and sebaceous glands, nerve formations of the skin explain the peculiarity of the skin reaction. Fever of any origin is associated with the loss of nitrogen, sulfur, phosphorus, potassium, vitamins due to increased breakdown of tissue protein, atsorexia, dietary restrictions, and often as a result of diarrhea and vomiting.[14] During the period of recovery it is necessary to provide the body with a sufficient amount of protein, because at this time should be restored lost tissue protein. In this case, nitrogen balance is made positive, and quickly goes weight gain. In chronic diseases the need of the organism in protein is provided mainly at the expense of its own muscle protein, so the protein of food goes to restore the protein of vital organs, and growth is slowed down. If the high need for protein is not fully satisfied, there is a slowdown or stoppage of development, growth, falling body weight, there is hypoproteinemia, edema, decreases the content of enzymes in digestive juices, especially in the secretion of the pancreas. Liver obesity develops, deamination processes are disturbed. Experiments on 20-day-old white rats showed that the transition from a full-fledged diet, in which proteins accounted for 18% of the total caloric content, to a diet in which proteins accounted for 9 and 4.5% of the total caloric content, very quickly leads to a violation of deamination and urea formation. Corresponding to the fall of urea nitrogen in the urine, amino acid nitrogen increased. In healthy infants and hypotrophy in the urine is never found tyrosine, phenylalanine, as well as products of their incomplete oxidation. Accession to hypotrophy any nonsevere disease causes the appearance in the urine tyrosine (up to 30-35lsh%), products of its incomplete oxidation (up to 25-30 mg%), phenylpyro- grape acid (up to 200-230 mg%). Protein deficiency reduces the defenses of the org-ganism, limits immunogenesis, decreases phagocytic activity. In this connection infection easily occurs. In these conditions, respiratory and intestinal infections are especially frequent, further aggravating protein deficiency and often leading to lethal. Healing of wounds after surgeries, traumas is delayed, complications often occur. In the state of starvation (hypotrophy) in children there are changes in the nervous system, expressed in delayed development of its functions, in hyporeflexia, instability of conditioned reflexes, in weakness of the main nervous processes - excitation and inhibition, especially internal inhibition, in paradoxical. It is believed that on the changes in the protein pattern of blood in young rats under conditions of starvation are of great interest. During 3 months the animals received a diet containing 3.2% protein (with the protein content in the food of the control group-18%). This is due to the physiological function of globulins. Normal gamma globulins can fix viruses on themselves and neutralize them without being antibodies.It should be assumed that in children and adults normal globulins, on the one hand, play a role in the regulation of the internal environment of the organism, and on the other hand, perform a protective function, and this protective function should be considered only as “a specialized case of the use by the organism of the reactive function of globulins for fixation and neutralization of harmful agents. In the overwhelming majority of pathological processes are accompanied by hypoalbuminemia, which leads to a violation of oncotic pressure of plasma, since the albumin fraction contains glucoprotein, which is of great importance in maintaining the constancy of colloidal osmotic pressure of the internal environment of the organism. Albumin is rich in sulfhydryl groups (SH), which makes them very reactive - they easily bind with various substances, causing their transport. It was mentioned above that the ability to bind and transport hormones and other biologically active substances also possess globulins. In addition, globulins have an important role in immune processes. Some fractions of globulins, mainly gamma-globulins, are humoral antibodies. High dispersibility of albumin facilitates their penetration through cell membranes, and cells use amino acids of albumin to build their own structures. Getting into the mitochondrial apparatus, albumin binds substances that separate the co-relation of respiration and phosphorylation. In the characterization of nutritional disorders special consideration should be given to the excessive introduction of protein to newborns and premature infants. It is known that excess protein in the food creates conditions for the development of acidosis. Compensation for it occurs through the mobilization of calcium and phosphorus from the bones, as a result of which may disrupt the process of bone formation and the formation of static functions. Bone thinning due to calcium loss (demineralization) has been observed in rats on an exclusively protein diet. As a result of imperfect barrier function of the intestinal wall in small children can occur absorption of unsplit protein from the intestine. Protein overfeeding creates conditions conducive to enteral sensitization. The previous material shows that for the developing organism is dangerous as a lack of protein, and its excess. It is not necessary the amount of protein, optimal for each age. At the same time, the condition of the child should be taken into account. Thus, previous protein starvation requires increased introduction of protein, exceeding the norm of a healthy child. [13]A number of disorders of amino acid metabolism are hereditary and manifest themselves at an early age. These include: phenylketonuria, alkaptonuria, albinism, cystinuria, hepato-cerebral dystrophy (Wilson-Konovalov disease or hepato-tolenticular degeneration) and some others. Phenylketonuria, or phenylpyruvine oligophrenia, is an inherited anomaly of amino acid metabolism, transmitted by autosomal recessive type and phenotypically manifested in homozygotic children autosomal recessive type and phenotypically manifested in homozygotic children. At phenylpyrovinogradnoy oligophrenia abnormal gene causes in the liver of children absence or decrease in activity of the enzyme (phenylalanine

hydroxylase), necessary for transformation of phenylalanine in tyrosine. This reaction is carried out with the coenzyme NAD (nicotinamidadenine dinucleotide) and in the presence of oxygen. In the liver of healthy children phenylalanine is converted into tyrosine with the participation of two enzymes, in the disease, one of them, more often mentioned above, is absent. As a result, the conversion of phenylalanine into tyrosine is inhibited. Phenylalanine is overaminated with ketoglutaric and other acids, resulting in the formation of fetsilpyruvic acid and other products of its transformations. In the blood sharply increases the content of phenylalanine, large amounts of it appear in the urine along with . Phenylpyruvic acid, phenyl lactic and other acids. Phenylpyruvic acid-base acts toxically on the developing brain, which leads to microcephaly and mental retardation. Areas of demyelination are found in the brain. With tyrosine deficiency, thyroid hormones are not synthesized, the starting material for which are tyrosine, which causes hypothyroidism of the developing organism. The amount of serotonin in the blood decreases. In phenylketonuria, melanin synthesis is also impeded, as a result of which sick children usually have very light coloring of skin, hair and iris. In England, a method of early diagnosis of phenyl-ketonuria was developed. Commercially produced papers impregnated with the reagent “Reshz^eh”, laid in the diaper of the newborn. If the baby's urine contains phenylpyruvic acid, the paper is colored brown-green. Early detection of metabolic anomaly allows you to protect the brain of the child by prescribing a special diet with a minimum content of phenylalanine. For this purpose, artificial milk is used, consisting of cabbage juice, soy protein, peanut oil, sugar, salt, Irish moss (as a bleach). In the Soviet Union also has a diagnosticum for phenylketonuria, but its insufficient sensitivity allows to diagnose the disease not earlier than three months of age of the child, when the violation of amino acid metabolism is sharply expressed and with urine excreted a significant amount of phenylpyruvic acid. To correct metabolism, a synthetic diet including a set of amino acids without phenylalanine is used. After several years of the child's stay on this diet, the cells of the nervous system become resistant to the action of phenylpyruvic acid and the diet is canceled. Alkaptonuria - a syndrome resulting from an inherited defect in amino acid metabolism and transmitted by thein an autosomal recessive pattern. Patients inherit an inability to synthesize the enzyme homogentinase in the liver, which is necessary for the conversion of homogentisic acid (an intermediate product of tyrosine metabolism). As a result, homogentisic acid accumulates in the liver, due to which the oxidation of all previous stages of tyrosine oxidation is inhibited; oxyphenylpyruvic acid, oxyphenyl lactic acid, tyrosine and other amino acids appear in the urine along with homogentisic acid. A large number of amino acids in the urine indicates a violation of the balance between the synthesis and their decomposition. Homogentisic acid, polymerizing in the presence of oxygen, forms a dark pigment alkapton, deposited in connective tissue, joints, especially in their cartilage, skin. Later, as a result of calcium deposition in the affected cartilage of the joints, arthritis develops.[16] Albinism is caused by the absence of tyrosinase in melanocytes, without which tyrosine cannot be converted into melanin. Tyrosine transformations proceed in two ways. Firstly, tyrosine is used to synthesize thyroxine and adrenaline, and secondly, tyrosine serves as a source of melanin formation. Lack of melanin in the skin does not cause pathological phenomena, but its low content in the iris increases the sensitivity of the eye to light, and as a result, rapidly develops a decrease in visual acuity. Albinism is inherited by autosomal recessive type, but significantly more common is partial albinism, transmitted by autosomal dominant type. Cystinuria is manifested by the fact that persons suffering from this anomaly of metabolism, with urine excreted large amounts of cystine (up to 1000 mg) and to a lesser extent - lysine, arginine and ornithine. The content of these amino acids in the blood is normal, and their increased excretion with urine depends on the absence of special enzyme systems in the renal tubule epithelium, as a result of which there is absolutely no reabsorption of cystine and 50% reduced reabsorption of lysine, arginine and ornithine. Since cystine can only be present in solution at concentrations not exceeding 400 mg/L, its higher content in the urine leads to precipitation and the formation of urinary cystine stones. Hepato-cerebral dystrophy (Wilson-Conoval disease, or hepato-lenticular degeneration) is caused by hereditary inherited decrease or perversion of synthesis of a certain fraction of alpha-2-globulin, called ceruloplasmin, which is a carrier of copper. In normal conditions, copper, coming with food in the gastrointestinal tract, absorbed into the blood, enters into an unstable connection with albumin, in the liver, copper is detached from albumin and, joining strong bonds with ceruloplasmin, circulates in the blood in conjunction with it. In hepato-lenticular degeneration as a result of reduced synthesis of ceruloplasmin, copper is easily detached from albumin, deposited in excessive amounts in tissues and excreted with urine. In the kidneys, copper forms complexes with amino acids, as a result of which their reabsorption becomes impossible and aminoaciduria develops. Deposition of copper occurs mainly in the liver, brain, kidneys, cornea, which leads to the development of cirrhosis of the liver, to degenerative changes in the tubules of the kidneys. Significantly reduced bioelectric activity of the brain, develop mental disorders, seizures, disorders of motor and coordination acts. The disease is inherited by autosomal recessive type.There is a group of inherited disorders of protein synthesis, which includes “double” albuminemia, characterized by the appearance of prealbumin and postalbumin on the electrophoregram.[3]

Carbohydrate Metabolism

In children in the first hours after birth, the content of sugar in the blood is the same as in the mother. However, later they develop hypoglycemia (on average up to 50-75 mg%), which should be explained by insufficient enzymatic activity of the liver. The large energy requirement of young infants causes glucose tolerance. A glucose load twice that of adults does not produce significant hyperglycemia. Sugar curve always has a flattened character. Hypoglycemia in newborns is considered physiologic and persists for several weeks. There is a hypothesis that the basis of hypoglycemia is a large consumption of sugar by the body during the period of adaptation to postnatal life, also hypoglycemia may depend on the high tone of skeletal muscle and a large production of insulin. The abundance of islet tissue, high resistance of beta-cells and high regenerative capacity of the islet apparatus in the newborn speaks in favor of enhanced insulin production.[4]The hypothesis explaining hypoglycemia in newborns and children of the first weeks of life by high glucose utilization by peripheral tissues is not recognized by all and is even considered doubtful. Scientists consider physiologic hypoglycemia of newborns as an expression of immaturity of glycogenolytic function of the liver. Even greater resistance to hypoglycemia is found in preterm infants. This resistance is parallel to resistance to hypoxia and, apparently, depends on insufficient development of the cerebral cortex and associated low metabolism of brain substances. Only significant shifts in blood sugar content (below 50-60 mg%) cause severe functional and morphological changes in the brain, slowing the formation of capillaries, differentiation of neurons. Prolonged hypoglycemia (sugar level below 60-50 mg%) leads to the development of cardiomegaly in the child, caused by additional work of the heart in violation of the energy supply of the organism. Insufficiency of regulatory mechanisms, in particular hormonal, makes it impossible to include neoglycogenesis, and therefore hypoglycemia, especially at a sugar content of 25-20 mg%, can cause paralysis of the respiratory center. It is characteristic that the paralysis occurs without the preceding stage of its excitation. A similar mechanism underlies hypoglycemia in Addison's disease, pituitary tumor. Changes in the brain are similar to those in intrauterine asphyxia. Hyperglycemia in early childhood occurs as a pro- manifestation of the reaction to extreme

influences, as a result of excitation of the sympathetic-adrenal system (in severe infectious diseases, shock caused by trauma, burns, hyperthyroidism, diabetes, etc.).Excessive content of carbohydrates in the food of the child, unbalanced with other components of the diet, can lead to disorders of phosphorus-calcium metabolism and contribute to the development of rickets. This depends on the fact that the absorption of sugar in the intestine is associated with the processes of phosphorylation, requiring inorganic phosphorus, and with a lack of the latter it is mobilized from the bones. "Children rarely have diabetes mellitus. According to \\\Tn1e (1964), among all patients with diabetes persons under 15 years of age are 5%, and among children, one sick child is yah 2500 healthy. Diabetes is often a hereditary heterogeneous disease, as evidenced by the high frequency of disease in both partners of monozygotic twins (58%), compared with 13% of disease in both partners of dizygotic twins. Diabetes is a polygenic disease. Apparently, different causes can cause mutation of different genes, as a result of which the disease occurs.[15]Diabetes mellitus in childhood flows severely and the younger the child is, the more severe the prognosis. Diabetes in children is characterized by rapidly developing emaciation, tendency to develop ketosis and diabetic coma. High energy demand in a small child is provided by intensive combustion of fats and carbohydrates. With a lack of the latter or in violation of their oxidation quickly increases the amount of ketone bodies. This explains the tendency to ketosis, noted in early childhood. The tendency to develop coma depends on the instability of hormonal regulation of metabolism in children and easily occurring disorders of balance of the autonomic nervous system. Even a mild infectious disease, mental trauma, food error, severe physical stress can cause the development of coma in children with diabetes. Often the course of diabetes is complicated by infectious diseases, tonsillitis, reducing the effect of insulin, increasing metabolism and contributing to the development of coma.In children, diabetes mellitus may occur in a latent-compensated form. With repeated determinations of blood sugar content, produced throughout the day, children show sharp fluctuations in its level from hypo- to hyperglycemia, not associated with food intake. Introduction of insulin is usually produced in certain hours, which may not meet the needs of metabolism, very unstable in childhood. Therefore, the child often has an absolute or relative insufficiency of insulin, as a result, there is a mobilization of carbohydrates, strengthening the processes of neoglucogenesis, and hypoglycemia is shifted by periods of hyperglycemia and ketonemia. Thus, not corresponding to the needs of the body supply of insulin, administered for therapeutic purposes, leads to hidden decompensation of the disease. In children in the first years of life, diabetes mellitus occurs with significant liver enlargement, growth retardation and delayed sexual development - Moriak's syndrome.[8]There is a group of hereditary diseases, which are based on the disorder of glycogen metabolism due to genotypically determined absence of any enzyme: glucose-6-phos- phatase, acid alpha-glucosidase, amylo-1-6-glucosidase, amylo-(1,4-1,6)-transglucosidase, muscle phosphorylase, liver phospho-rylase.[10] The absence of one of these enzymes, violating the exchange of glycogen, leads to its accumulation in the cells of the liver, kidneys, myocardium, lungs, spleen, causing the enlargement of these organs. At the same time, the content of sugar in the blood is reduced, due to which hypoglycemic convulsions may occur. Glucose-6-phosphatase is absent in the liver of mammalian embryo and appears only at the end of the intrauterine period. Insufficient supply of glucose to the tissues of the newborn leads to growth retardation, muscle weakness. The disease is transmitted, apparently, in a polygenic recessive manner. Currently described 12-14 forms of glycogenosis, of which the most studied is glycogen disease (type I), or Hirke's disease. The disease may appear soon after birth and, in addition to the above signs, is characterized by acidosis caused by the accumulation of lactic, pyruvic and fatty acids in the blood, hypercholesterolemia, osteoporosis. In Girke's disease in the liver lacks glucose-6-phosphatase, resulting in blocked conversion of glucose-6-phosphate to glucose, and although the liver and other organs are saturated with glycogen, the blood glucose content is low and tissues lack it. In the body of healthy normal-fed children after the introduction of adrenaline or glucagon in 10 to 20 min depolymerization of glycogen occurs.20 min depolymerization of glycogen to glucose-1-phosphate, glucose-6-phosphate occurs, and after dephosphorylation free glucose and inorganic phosphorus enter the blood. In patients with glycogenosis this reaction is not carried out due to the absence of glucose-6-phosphatase, and therefore the administration of adrenaline or glucagon is not accompanied by hyperglycemia. The liver cells overfilled with glycogen are not able to process food carbohydrates into glycogen, which leads to sharp fluctuations in blood sugar levels during the day (60 me% in the morning and 176 mg°/o in the middle of the day).Galactosemia is a severe inherited abnormality of carbohydrate metabolism. It is inherited by the autosomal recessive type. The body does not form the enzyme galactosauridine transferase, which is necessary for the conversion of galactose into glucose. As a result, galactose and galactose-1-phosphate accumulate in the blood, hypoglycemia and galactosuria develop. Galactose-1-phosphate acts toxically, inhibiting the activity of a number of enzymes, such as phosphoglucomutase, glucose-6-phosphate dehydrogenase, which disrupts glucose metabolism. Develop obesity of the liver with subsequent cirrhosis, mental retardation, cataracts, leading to blindness. Early diagnosis of this disease is very important, as timely elimination of lactose and galactose from food prevents its occurrence in a child. The disease appears in infants when feeding them with milk.To correct the hereditary defect - lack of enzyme - the method of genetic construction is promising. For this purpose, the cells of the organism suffering from galactosemia are brought into contact with a phage or a virus that previously parasitized E. coli and in the process of reproduction assimilated a certain amount of genetic material of E. coli, namely, genes encoding the synthesis of galactose phosphofosfouridine transferase. In this way, it is possible to achieve assimilation of inherited information of phages or viruses encoding the synthesis of the necessary enzyme by the cells of the organism. After the experiments of scientists, who managed to hybridize DNA, the possibility of hybridization of nucleic acids became obvious.A number of other anomalies of carbohydrate metabolism are known (pentosuria, levulesuria, fructosuria), also caused by hereditary disorder of synthesis of a particular enzyme.[9].

Lipoid Metabolism

The level of total blood cholesterol, the ratio of esters and free fraction of cholesterol is one of the constants of the organism. For a newborn child is characterized by a low content of cholesterol in the blood - 65 mg%, which is 2 times lower than in the adult body.in the adult organism. Twice lower and the coefficient of Ephospholipids

equal to 0.56 (adult 0.98). In children's bodies are often observed disorders of regulation of cholesterol levels in the blood, manifested by hypercholesterolemia or hypocholesterolemia. In the pathogenesis of diseases such as Niemann-Pick disease, progressive lipodystrophy, lipoidosis, Gaucher disease, lipoid nephrosis, lies a violation of the regulation of lipoid metabolism, proceeding with high hypercholesterolemia (up to 800 mg%) and with the deposition of cholesterol esters in reticuloendothelial cells. Leukemia, severe anemia, hepatitis in children with hypocholesterolemia, which is considered as an unfavorable factor. During remission in leukemia blood cholesterol level rises, the disappearance of anemia is characterized by a return of cholesterol level to the original, ie, to a higher level. It is also known that the period of antibody production coincides in time with an increase in the blood cholesterol level. Scientists obtained the death of animals by

administration of non-denatured DNA, which caused a persistent blockade of endogenous cholesterol synthesis, and thereby prolonged hypocholesterolemia.[1]. The need of children's body in cholesterol is very great, as cholesterol is a plastic substance, a necessary structural component of the cell. It is included as an obligatory element in the composition of cellular and subcellular membranes and thus affects the passive and active transport of substances across the membrane. It is an obligatory component of mitochondria, where two vital processes of the cell intersect: energy and genetic. As a highly hydrophilic substance, cholesterol is involved in osmotic processes. It is contained in large quantities in brain tissue (in white matter - f%|, in gray matter - 2.3%), being a part of myelin, provides insulating properties of central nerve fibers and thereby directional conduction of impulses. It is abundant in the cortex of adrenal glands. Free hydroxyl groups of the cholesterol molecule bind substances such as saponins, performing a detoxification function. The presence of cholesterol in the skin indicates its relation to the function of the keratinous substance of the skin. The body covers its needs in cholesterol through the synthesis of endogenous cholesterol, which is 2/3 of all cholesterol in the body. Only 1/3 of all cholesterol of the body comes with food. These quantitative ratios already allow us to confirm the position about the greater importance of endogenous cholesterol in the vital activity of the organism, especially in children, in which growth, body weight increase, and the processes of diff ferentiation of cell populations actively take place.[6]In the pathogenesis of disorders of cholesterol metabolism in children and in the adult organism is important violation of metabolism of endogenous cholesterol. Scientists by their studies showed in the experiment that at early stages of ontogenesis there is a very active synthesis of cholesterol, its rapid accumulation in brain tissue, even when animals are on a low-calorie diet. This emphasizes the importance of cholesterol in the plastic processes, so strongly expressed in the growing body. Comparing the amount of cholesterol, introduced into the body, with the amount excreted in the form of bile acids, the authors found that in animals of early age cholesterol is excreted 2-4 times more compared to the introduced. These data also speak about active synthesis of endogenous cholesterol occurring at early stages of ontogenesis. Load of exogenous cholesterol at this time causes an increase in its level in the blood due to the imperfection of liver function on esterification, oxidation and excretion of cholesterol, which is subjected to decomposition in the liver and removed from the body in the form of bile acids. Violation of cholesterol metabolism can occur in the fetus at exogenous and endogenous cholesterolemia of the maternal organism. The authors found that in this case the fetus has hypercholesterolemia, increased beta-lipoproteins, increased content of total cholesterol in its tissues, obesity of the liver, and sometimes myocardium. Studies scientists with the use of labeled cholesterol ('C14) allowed to establish a violation of the placental barrier for cholesterol in connection with the sharp obesity of the fetal part of the placenta, the chorionic villi of which contained the amount of cholesterol, 6 times higher than its content in the norm. Scientists found in the villi of the fetal part of the placenta a high concentration of free fatty acids and phospholipids. Since their content in the villi was higher than in the blood of the maternal organism and in the blood of the umbilical cord, the authors believe that the synthesis of fatty acids and phospholipids occurs indirectly in the villi of the placenta.[17]

Fat Metabolism

The main energy material in the child's body are triglycerides. For fat oxidation must contain a sufficient amount of carbohydrates in the food to avoid the development of ketosis. Feeding mixtures, poor in fat, leads to a lack of fat-soluble vitamins. In a child's body the amount of absorbed in the intestine of dietary fat is less than in an adult. The smaller the child, the higher the percentage of unused fat. In premature infants, fat absorption only approaches adult levels by the end of the year. Children from 3 to 10 years of age per day should receive 25-30 g of fats. The need of a newborn child in fats is not precisely established. The subcutaneous fat layer in newborns is poorly developed, and the fat composition is richer in fatty acids with a high melting point (stearic and palmitic).At the age of 2 to 10 years, children, regardless of the composition of food, easily give ketosis. Ketone bodies are formed in the liver, and their decay is accomplished mainly in tissues and is accompanied by the release of energy. Excitement, fatigue, infectious diseases in combination with ketogenic food quickly lead to ketosis, which is very favorable to the instability of carbohydrate metabolism. In infancy, ketonuria is a rare phenomenon. This is due to the characteristics of the renal barrier. Only at a high degree of keto- nemia ketone bodies appear in the urine.[5]In children there is an imperfect adaptation to alimentary load of fats, which is associated with insufficiency of lipolytic enzymes. Oserved a similar reaction in early rats.[19]

Water-Salt Metabolism

In the pathology of early childhood play a major role in disorders of water-salt metabolism. The younger the organism, the easier it is to develop hyperhydration, edema, hypohydration (exci- kosis). This is due to the peculiarities of water-salt metabolism, characteristic of early age, easy vulnerability of mechanisms that regulate water and salt homeostasis, the consequence of which are diseases of infants, often accompanied by diarrhea, vomiting, malnutrition. The younger the organism, the richer it is with water. In premature babies the water content in the organism is higher than in premature babies. In children with a birth weight of 1500-2500 g, the total water content is 81-85% of body weight. This is due to the fact that a small child has less cell mass, and therefore" and the content of intracellular fluid, and relatively more of its surface - skin, rich in extracellular fluid. At the early stages of ontogenesis extraclotid fluid has great mobility, which depends on the small development of connective tissue structures, due to which water is not fixed in them and easily moves between interstitial and intravascular. Therefore, infants very easily lose water and also quickly restore its volume under creation of appropriate conditions.[2]. The high percentage of water content in the body of a small child or animal gives the impression that it is very rich in water and, therefore, should be resistant to dehydration. However, this is not the case at all. The younger the age of the child, the greater the surface area per unit body weight, and therefore, the higher his metabolism and extrarenal water loss. Thus, at rest by means of transpiration, an infant loses water through the skin in the amount of 1 ml/kg/h, while an adult loses 0.5 ml/kg/h.For estimation of indicators of water metabolism of a child it is important to make calculation per unit of weight and body surface. It turns out that the volume of extracellular fluid per 1 kg of weight in a child is more significant than in an adult, and when calculated per unit of body surface it is less than in an adult. By the way, the same discrepancies are found in relation to the minute volume of the heart, systolic volume, oxygen consumption and others. At an early age, the mechanisms that regulate the constancy of the volume and composition of fluid spaces of the body, has not yet reached the final formation™. The sensitivity of volum and osmoreceptors of the vascular system and tissues in small children is low. In newborn children of the first months of life osmoreceptors supraoptic and paraventricular nuclei of the hypothalamic region do not yet have a pronounced osmoregulatory function. The concentrating mechanism of the kidneys does not work (see section "Kidneys"). This causes physiological polyuria and hyposthenuria, characteristic of the infant. During the first year of life, despite the incompletely formed excretory apparatus of the kidneys, the degree of their development corresponds to the needs of this age period and with the maintenance of optimal living conditions provides homeostasis. However, violations of nutrition,

temperature regime lead to homeostasis violation. Thus, excessive introduction of protein, salts in the body leads to azotemia, to hypersalemia. In newborns water excretion from the body occurs at a slower rate than in older age. Thus, a newborn baby during 2 h removes only 10% of the water drunk, while 3-month-old for the same time removes all the water received. Newborn children and animals cannot quickly remove excess salt, as a consequence of which they easily suffer from osmotic disequilibrium and secondary fluid retention. Immaturity of regulatory mechanisms that provide stability of water-salt metabolism, is the basis of its frequent disorders in early childhood. Dehydration (ecicosis), or, in other words, dehydration, can be caused in young children by a number of reasons: diarrhea, vomiting, starvation, abundant perspiration, hyperventilation syndrome, gatekeeper stenosis, primary circulatory disorders, adrenal insufficiency and some other rare causes. Dehydration in infants with insufficient fluid intake develops earlier than in adults, as the child loses large amounts of fluid with urine and perspiration with exhaled air. The most common cause of dehydration in children are acute alimentary and infectious lesions of the gastrointestinal tract, causing diarrhea, vomiting and giving a picture of severe dehydration. This condition is called toxic dyspepsia (for more details, see the section “Digestive system”). Sometimes this toxic state occurs in acute respiratory diseases, otitis media, pneumonia.[18]. When calculated per unit of surface area of the infant has a minute volume of the heart is almost the same as an adult, with half the volume of circulating blood. Dehydration in toxicosis, causing hypovolemia and blood clotting, creates extremely unfavorable conditions for blood circulation, the consequence of which may be the development of collapse. In severe toxic dyspepsia as a result of dehydration plasma volume can fall by almost [7]. This leads to a decrease in the minute volume of the heart. Arterial pressure decreases, almost 4 times slower blood flow velocity, blood oxygen saturation falls, arterio-venous oxygen difference increases. Due to a decrease in the minute volume of the heart and the associated impairment of blood supply to the kidneys, urea clearance, which is low in a healthy child, falls even lower in toxicosis, and extrarenal uremia develops. Formed in the intestine in infectious lesions of its toxic products and products of incomplete oxidation, arising in tissues as a result of impaired blood circulation, increase the permeability of the vascular wal . Scientists found desolation of peripheral vessels and accumulation of blood in the vessels of the abdominal cavity. It is believed that the state of capillaries in children with dehydration caused by toxicosis, observed spasm of precapillaries and venules, interspersed with atonic expansion of them. Disturbance of microcirculation leads to hypoxia of a number of organs, to the occurrence of dystrophic changes in them. So, in most cases of toxic dyspepsia they are found in the myocardium, in the adrenal glands, in the liver. The consequence is a violation of detoxification, urea-forming and other functions of the liver [12-20]. As noted, dehydration in children can be observed in hyperventilation syndrome, which occurs with fever in the case of diseases such as influenza, pneumonia, sepsis, encephalitis. Hyperventilation syndrome is characterized by shortness of breath. Exhaled air is 80% saturated with water vapor, and with increased frequency and deepening of breathing (hyperventilation) there is a loss of water through the lungs without loss of salts. In extreme cases, water loss by this way can reach 20 g/kg/day. Hyperventilation occurs reflexively or has a central origin, such as in encephalitis. Fluid loss leads to blood clotting, hypovolemia, the consequence of which may be collapse. Despite hyperventilation and a fall in the content of increased lota in the blood, pH may not change significantly, such blood is deprived of the formation of acids in tissues experiencing' Polynatal starvation. Hyperchloremia occurs, the excretory capacity of the kidneys is impaired, resulting in azotemia, hyperphosphatemia [7]. Dehydration often occurs in children during starvation. Quantitative and qualitative lack of food, infectious diseases with lesions of the gastrointestinal tract in children of the first year of life lead to alimentary dystrophy. If there is no sharply expressed protein deficiency in the child's diet - edema does not develop and normal concentration of digestive juices in the intestine is preserved [5-9]. Protein insufficiency leads to hypoproteinemia, to a decrease in the colloid-osmotic pressure of blood, to edema. Regularly decreases the concentration of enzymes in duodenal juice due to the lack of a number of essential amino acids, which leads to diarrhea, liver obesity develops. With the loss of body weight, the ratio of extracellular fluid volume to body weight increases, although the absolute amount of it may remain unchanged. In the pathogenesis of starvation edema, along with a drop in colloid osmotic pressure of blood, is important hypovolemia, which excites volum of receptors, leading to increased secretion of aldosterone. It is possible that hypovolemia causes the secretion of renin - angiotensin, and the latter, affecting the tubular zone of the adrenal cortex, increases the production of aldosterone. Aldosterone increases sodium reabsorption in renal tubules, which contributes to hypertension of extracellular fluid. This, in turn, causes an increase in the secretion of antidiuretic hormone. Thus, there is a retention of sodium and water in the body [4-8]. When starvation is possible development of starvation edema (hyperhyderation). Their pathogenesis is not finally clarified. Both extracellular and intracellular fluid were found to be increased, with cellular sodium concentration increased and plasma sodium concentration decreased. Starving adults were much less likely to develop hyponatremia, which is related to the high intake of table salt in adults. The cellular potassium content in children with starvation edema was very low. If the sodium content in extracellular fluid in starvation edema did not fall, the volume of only extracellular fluid increased [14-21]. Dehydration is seen in gatekeeper stenosis, which occurs early in life, and is manifested by indomitable vomiting. Starvation resulting from vomiting leads to breakdown of the body's own tissues. The body loses potassium, with vomit masses and due to the exchange of intracellular potassium for extracellular sodium. Dehydration develops, but it progresses more slowly than exhaustion from starvation. In advanced cases, death comes from hypoglycemia. Dehydration occurs in congenital adrenocortical syndrome with salt loss. In this case, it is a hereditary disorder of corticosteroid metabolism. The disease is accompanied by vomiting, and electrolytes are lost both with gastric juice and by excretion of salts by the kidneys, leading to dehydration, hyposalemia and hyperkalemia.

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