

Maxillofacial Aspects of Metabolic Disorders

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ABSTRACT

Metabolism could be an advanced method that involves a series of chemical reactions within the flesh. Alterations in these metabolic processes represent the disturbances of metabolism inflicting metabolic disorders. The breakdown of sugar by oral microorganisms has been the topic of diverse investigations. This review discusses the oral aspects of metabolic diseases.

Keywords: Metabolism; Oral flora; Metabolic disorder

INTRODUCTION

Metabolism

- Duncan outlined metabolism as “the accumulation of tissue activity as thought-about in terms of chemical science changes related to and controlled by the provision, utilization and disposal of macromolecule, fat, macromolecule, vitamins, minerals, water and also the influences that the endocrines exert on these processes”. Metabolism could be a advanced method that involves a series of chemical reactions within the body. Some reactions turn out the energy that is keep within the type of nucleotide whereas alternative reactions consume energy to manufacture advanced compounds. the method of metabolism includes two varieties of mechanisms, i.e, destructive metabolism and organic process [1].
- Catabolism could be a method that involves unharness of energy by breaking advanced organic compounds into straightforward molecules. Thus, during this method energy is been made.
- Anabolism could be a method that needs energy for synthesis of advanced compounds from little molecules. during this method energy is been utilised.
- Alterations in these metabolic processes represent the disturbances of metabolism inflicting metabolic disorders. The breakdown of macromolecule by the oral microorganisms has been the topic of diverse investigations [2]. it's believed by many who the intraoral production of acids from macromolecule, arising from incomplete reaction, could be a think about the assembly of decay, dental medicine diseases, oral reek, bone loss and alternative associated metabolic diseases [3].
 - Types of Metabolic Disorders [4,5]
 - Disturbances in macromolecule metabolism
 - Disturbances in lipide metabolism
 - Disturbances in macromolecule metabolism
 - Disturbances in internal secretion metabolism
 - Lysosomal storage disorders
 - Mitochondrial storage disorders

Disturbances of protein metabolism

Amyloidosis: it's a rare unwellness that results from accumulation of unsuitably doubled proteins. These misfolded proteins area unit referred to as amyloids. once proteins that area unit usually soluble in water fold to become amyloids, they become insoluble and deposit in organs or tissues, disrupting traditional perform.

Protein energy deficiency disease: PEM includes malnutrition (protein malnutrition predominant), malnutrition (deficiency in calorie intake), Marasmic malnutrition (marked macromolecule deficiency and marked calorie insufficiency signs gift, typically brought up because the most severe type of malnutrition) PEM is fairly common worldwide in each youngsters and adults and accounts for six million deaths annually. PEM could also be secondary to different conditions like chronic urinary organ unwellness or cancer infirmity during which macromolecule energy wasting could occur.

Porphyria: The porphyrias area unit a bunch of rare diseases during which chemical substances referred to as porphyrins accumulate. The body needs porphyrins to provide haematin, that carries chemical element within the blood; however, within the porphyrias, there's a deficiency (inherited or acquired) of the enzymes that rework the assorted porphyrins into others, resulting in abnormally high levels of 1 or additional of those substances. This manifests with either medical specialty complications or skin issues or sometimes each.

Erythropoietic uroporphyrin: it's caused by genetic defects that cause deficiency of the protein uroporphyrinogen III cosynthase (UROS). The unwellness is characterized by extreme sensitiveness (abnormal body covering reaction to sunlight) which may leave severe scarring, blister formation and therefore the loss of digits or different options. broken skin will become infected, resulting in any sphaelus and deformities. The face, hands and arms area unit the foremost considerably affected as they're oftentimes exposed; typically presenting as severe disfiguration.

Disturbances in saccharide metabolism [6]

Mucopolysacchridoses result from abnormal degradation of glycosaminoglycans. Disorders in saccharide metabolism embrace Mucopolysacchridoses

MPS sort I includes twirler and Scheie syndrome: it's a cutaneous condition characterised by gentle retardation and tissue layer vapour

MPS sort II includes Hunter syndrome: (mild to severe form) it's a lysosomal storage illness caused by a deficient or absent catalyst, iduronate-2-sulfatase (I2S). The accumulated substrates in Hunter syndrome square measure heparan sulphate and dermatan sulphate. The syndrome has X-linked recessive inheritance.

MPS sort III includes Sanfillippo syndrome: It is a metabolism disorder passed down through families. It makes the body unable to properly break down long chains of sugar molecules known as glycosaminoglycans (formerly known as mucopolysaccharides). The syndrome belongs to a gaggle of diseases known as mucopolysaccharidoses (MPS).

MPS sort IV includes Morquio syndrome: it's AN chromosome recessive polysaccharide storage illness sometimes heritable. it's a rare form of anomaly with serious consequences like vas and ophthalmic defects.

MPS sort VIa includes Maroteaux- Lamy syndrome (classic form) MPS sort VIb includes Maroteaux- Lamy syndrome (mild form): it's a kind of hereditary disease caused by a deficiency in arylsulfatase B (ARSB). Symptoms embody medical specialty complications like clouded corneas, deafness, thickening of the dura mater and pain caused by compressed or traumatized nerves and nerve roots.

MPS sort VII includes beta glucuronidase deficiency: it's a awfully rare congenital disease with high fatality rate. birth defect in multiple organs with gender predilection for males.

Disturbances in macromolecule metabolism [7]

Gaucher's disease: it's a inherited disorder during which glucosylceramide accumulate in cells and bound organs. The disorder is characterised by bruising, fatigue, anaemia, low blood platelets, and enlargement of the liver and spleen. it's caused by a hereditary deficiency of the catalyst glucocerebrosidase.

Niemann-Pick disease: it's one in all a gaggle of lysosomal storage diseases that have an effect on metabolism which square measure caused by genetic

mutations. Enlargement of the liver and spleen (hepatosplenomegaly) might cause reduced appetency, abdominal distension, and pain.

Letterer-Siwe disease: it's the acute disseminated multisystem kind of Langerhans cell blood disorder characterized by proliferation of nonlipid histiocytes within the innards and bones. Clinical options embody a range of skin lesions,

osteolytic lesions, pathology, hepatosplenomegaly, pulmonic infiltration, spiking fever, anaemia, blood disorder, articulator dysplasia, animal tissue inflammation, loss of teeth, otitis, hemorrhages, failure to thrive, cachexia.

Disturbances in secretion metabolism [8]

Pituitary hormone: Hypopituitarism: Symptoms embody fatigue, low force per unit area, weight loss, weakness, depression, nausea, or projection, constipation, weight gain, sensitivity to cold, attenuated energy, and muscle weakness or aching. In women, symptoms embody irregular or stopped emission periods and sterility. Hyperpituitarism: Symptoms includes headaches, visual disturbance, and growth failure, weight gain. hairiness and premature start might occur in prepubescent kids. time of life arrest, acne, fatigue, and depression are common.

Thyroid hormone: Hyperthyroidism includes irritability, muscle weakness, sleeping issues, a quick heartbeat, poor tolerance of warmth, diarrhea, enlargement of the thyroid, exophthalmos and weight loss. Hypothyroidism includes poor ability to tolerate cold, a sense of temporary state, and weight gain. In kids, glandular disorder ends up in delays in growth and intellectual development that is termed hypothyroidism.

Parathyroid hormone: glandular disorder includes depression, fatigue, polydipsia and kidney disease, feeling sick and losing your appetency, muscle weakness, constipation, tummy pain, loss of concentration. glandular disorder includes muscle aches or cramps, tingling, burning, or symptom in fingertips, toes, and lips, muscle spasms, particularly round the mouth, uneven hair loss, dry skin, brittle nails, fatigue, anxiety or depression.

Lysosomal storage disorders [9,10]

Lysosomal storage disorders square measure a gaggle of roughly fifty rare heritable metabolic disorders results from defects in lysosomal perform. Lysosomes square measure sacs of enzymes among cells that digest massive molecules and pass the fragments on to alternative elements of the cell for utilization. This method needs many vital enzymes. If one in all these enzymes is flawed, due to a mutation, the big molecules accumulate among the cell, eventually killing it. The symptoms of lysosomal storage illness vary, reckoning on the actual disorder and alternative variables just like the age of onset, and might be gentle to severe. They embody organic process delay, movement disorders, seizures insanity, hearing impairment and/or vision defect, megalohepatic hypertrophy and internal organ issues, and bones that grow abnormally. Some examples square measure listed below.

Schindler illness/Kanzaki disease: Schindler disease results from the deficient activity of the catalyst alpha-N -acetylgalactosaminidase (alpha-galactosidase B), with the buildup of sialylated-asialo- glycopeptide and carbohydrate with alpha-N -acetylgalactosaminyl residues.

Faber disease: (Farber's lipogranulomatosis, ceramidase deficiency) is an especially rare (80 cases rumored worldwide to this day) chromosome recessive lysosomal storage illness marked by a deficiency within the catalyst ceramidase that causes AN accumulation offatty material lipids leading to abnormalities in the joints, liver, throat, tissues and central nervous system.

Krabbe disease: (globoid cell leukodystrophy) is a rare, often fatal degenerative disorder that affects the myelin sheath of the nervous system. It is a form of sphingolipidosis, as it involves dysfunctional metabolism of sphingolipids. This condition is inherited in an autosomal recessive pattern.

Tay-Sachs and Sandhoff diseases: is a rare autosomal recessive genetic disorder. It causes a progressive deterioration of nerve cells and of mental and physical talents that begins around six months elderly and frequently leads to death by the age of 4.

Pyknodysostosis: additionally celebrated as hereditary condition acro- osteolytica or Toulouse-Lautrec syndrome could be a rare chromosome recessive bone abnormalcy, defined by sclerosis and short stature.

Mitochondrial diseases [11]

Mitochondrial illness is also a cluster of disorders caused by dysfunctional mitochondria, the organelles that generate energy for the cell. Mitochondrial disorders is additionally caused by mutations, acquired or transmitted, in mitochondrial compound (mtDNA) or in nuclear genes that code for mitochondrial elements. they will even be the results of acquired mitochondrial pathology thanks to adverse effects of drugs, infections, or different environmental causes. Symptoms embody poor growth, loss of muscle coordination, muscle weakness, visual problems, hearing problems, learning disabilities, heart condition, disease, nephropathy, canal disorders, biological process disorders, medication problems, involuntary pathology and mental disease. samples of mitochondrial diseases include-

Luft disease: incorporates Hypermetabolism, with fever, heat intolerance, swarming perspiration, polyphagia, polydipsia, ragged-red fibers, and resting heart disease. Exercise intolerance with light weakness.

Leigh syndrome: incorporates acute sclerosing brain sickness} once ancient development the disease usually begins late among the initial year of life, although onset may occur in adulthood. A speedy decline in perform happens and is marked by seizures, altered states of consciousness, dementia, improvement failure.

Alpers disease: to boot referred to as as Progressive Infantile Poliodystrophy. Symptoms embody seizures, dementia, spasticity, blindness, liver pathology, and cerebral degeneration.

Pearson marrow syndrome: it is a mitochondrial illness defined by sideroblastic anaemia and secreter duct gland pathology. Clinical choices includes failure to thrive, monogenic disorder, muscle and medicine impairment, and, frequently, early death.

Wilson illness: is associate body recessive disorder throughout that copper accumulates in tissues this manifests as medication or medicine symptoms and disease. it's treated with medication that reduces copper absorption or removes the excess copper from the body, but usually a liver transplant is required.

Batten disease: it is a rare, fatal body recessive neurodegenerative disorder that begins in childhood. Early signs is additionally delicate temperament and behavior changes, slow learning or regression, repetitive speech or echolalia, clumsiness, or staggering. there's additionally retardation head growth among the infantile kind, poor circulation in lower extremities (legs and feet), belittled body fat and muscle mass, curvature of the spine, external respiration and/or breath-holding spells, teeth grinding, and constipation.

Menkes disease: it's a disorder that affects copper levels within the body resulting in copper deficiency. it's associate x joined recessive disorder, and is so significantly additional common in males: females need 2 defective alleles to develop the illness. Signs and symptoms of this disorder embody weak tone (hypotonia), droopy face expression, seizures, intellectual incapacity, blue sclerotic coat and organic process delay. The patients have brittle hair and metaphyseal widening. In rare cases, symptoms begin later in childhood and ar less severe. Affected infants is also born untimely.

CONCLUSION

Process of metabolism is incredibly advanced and integrated with multiple factors accountable for physiological condition. Oral microbic flora additionally plays a significant role during this method. Any quite amendment within the pathway from cellular to macroscopical level might make to variety of metabolic diseases related to native and general changes. This review enlightens numerous sorts of metabolic disorders and their associated oral conditions.

REFERENCES

- [1]. Ebenhöh O, Heinrich R (2001) organic process improvement of metabolic pathways. Theoretical reconstruction of the ratio of nucleotide and NADH manufacturing systems. Bull mathematics Biol
- [2]. Takahashi N, Sato T (2002) Dipeptide utilization by the dental medicine pathogens Porphyromonas gingivalis, Prevotella intermedia, Prevotella nigrescens and Fusobacterium nucleatum. Oral Microbiol Immunol
- [3]. Niederman R, Zhang J, Kashket S (1997) Short-chain carboxylic-acid- aroused, PMN-mediated animal tissue inflammation. Crit Rev Oral Biol Master of Education
- [4]. Takahashi N, Saito K, Schachtele CF, Yamada T (1997) Acid tolerance of growth and neutralizing activity of Porphyromonas gingivalis, Prevotella intermedia and Fusobacterium nucleatum. Oral Microbiol Immunol
- [5]. Mayanagi G, SatoT, Shimauchi H, Takahashi N (2004) Detection frequency of disease-associated microorganism by enzyme chain reaction in subgingival and supragingival plaque of subjects with periodontitis and healthy subjects. Oral Microbiol Immunol.