

* *Degradation of purine nucleotides :*

- occur in the small intestine .
- uric acid is the end product .
- mammals other than primates oxidize uric acid to allantoin .

* *Degradation of dietary nucleic acid in the small intestine :*

- DNA , RNA> denatured by low PH
- denatured nucleic acid ---- (nucleases) ----→ oligonucleotide
- oligonucleotide -- (pancreatic phosphodiesterases)-→- 3 & 5 mononucleotide
- nucleotides --- (nucleotidases remove P group) -→ nucleoside .
- * NOTE : here nucleoside may be absorbed by the intestinal mucosal cell , or be further degraded to free bases before uptake .
- nucleoside -----(nucleosidase) -----→ dietary purine & pyrimidine .
- dietary purine & pyrimidine have two ways :
 - 1) converted to uric acid by intestinal mucosal cell .(major)
 - 2) synthesis of nucleic acid . (minor)

* *formation of uric acid : (only point 1 ,5 in the book):*

- an amino group is removed from AMP to produce IMP . (AMP deaminase)
- an amino group is removed from adenosine to produce inosine (adenosine deaminase)
- NOTE : inosine also called (hypoxanthine ribose)
- Xanthine oxidase oxidize :
 - a) hypoxanthine to xanthine
 - b) xanthine to uric acid

* *Gout disease (will be studied later on practical class)*

***ADENOSINE DEAMINASE (ADA) DEFICIENCY : (MCQ) (MCQ)**

- in cytosol of all cells especially lymphocyte .
- autosomal recessive disorder .
- accumulation of adenosine .
- so accumulation of ribonucleotide & deoxyribonucleotide .
- large dATP in red cell which inhibit ribonucleotide reductase , so inhibit DNA synthesis . (MCQ)
- In severe form , it causes severe combined immunodeficiency Disease (SCID), involving lack of both T , B cells . (MCQ)
- children die before two years of age .
- *Treatment :*
 - bone marrow replacement or enzyme replacement .
 - is the first genetic disease treated by gene therapy .

Adenosine deaminase deficiency is:

- X-linked syndrome
- Characterized by the accumulation of dGTP in red cells
- manifested by mental retardation & self mutilation.
- Characterized by severe combined immunodeficiency (SCID). (answer)
- Associated with hyperuricemia.

Severe combined immunodeficiency syndrome is:

- associated with inhibition of ribonucleotide reductase.
- associated with hyperuricemia.
- associated with the accumulation of DGTP in red cells.
- characterized by T-dysfunction only.
- due to purine nucleophosphorylase deficiency.

تمت المحاضرة الثالثة.....

Done by :

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