Adenosine Deaminase Deficiency

Adenosine deaminase deficiency is an autosomal recessive metabolic disorder that is one of the major causes of severe combined immunodeficiency (SCID). Adenosine deaminase is an enzyme involved in purine metabolism. It is used to break down adenosine to inosine for the turnover of nucleic acids for DNA synthesis and cell growth. This deficiency leads to an accumulation of deoxyadenosine which leads to a buildup of ATP. High ATP, which normally activates RNR, leads to high dATP, which inactivates the enzyme ribonucleotide reductase, which is a key enzyme in DNA synthesis. This in turn prevents DNA synthesis. Because developing T cells and B cells are extremely mitotically active, these cells are susceptible to cell death. Loss of B and T cell function causes severe immunodeficiency.



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Adenosine to Inosine
A-dentist-singing becoming N'SYNC
Adenosine deaminase is an enzyme involved in purine metabolism. It is used to breakdown adenosine to inosine for the turnover of nucleic acids for DNA synthesis and cell growth.

Excess ATP
Excess ATP-batteries
Adenosine deaminase deficiency leads to an accumulation of deoxyadenosine which leads to a buildup of ATP and dATP in cells.

Inhibition of Ribonucleotide Reductase
Red-bull Red-duck
High ATP, which normally activates RNR, leads to high dATP, which inactivates the enzyme ribonucleotide reductase, which is a key enzyme in DNA synthesis.

Prevents DNA Synthesis
DNA Double-helix
High ATP, which normally activates RNR, leads to high dATP. This inactivates the enzyme ribonucleotide reductase, which is a key enzyme in DNA synthesis. Thus, DNA synthesis is prevented.

Decreases B Cells and T Cells
Down-arrows on Basketball and Tennis-ball
Developing B cells and T cells are extremely mitotically active and thus largely affected by the disruption in DNA synthesis. Lack of B and T cells results in high susceptibility to viral, fungal and bacterial infections.

Major cause of SCID
SCID marks
Adenosine deaminase deficiency accounts for about 15% of cases of severe combined immunodeficiency (SCID).

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