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9am to 5pm



Supporting those affected by  
Inherited Metabolic Disorders

# Beta-Aminoisobutyric Aciduria

Beta-Aminoisobutyric Aciduria is a defect of pyrimidine catabolism. Pyrimidines are one of two important families of compounds called nitrogenous bases. When paired with the other family (purines) they make up the building blocks of DNA and, when involved in the construction of RNA, pyrimidines are involved in the production of all the proteins in the body's cells. Pyrimidines are broken down in four steps each requiring a different enzyme. A defect in an enzyme called BAIBPAT causes a high excretion of Beta-Aminoisobutyric Acid in the urine. Beta-Aminoisobutyric Aciduria is benign and does not require treatment.

## Synonyms

Alternative names for this condition are:

- BAIBA
- Hyper-Beta-Aminoisobutyric Aciduria
- Urinary Excretion of Beta-Aminoisobutyric Acid

Further information is available from Climb.

## Disclaimer

[Please read our disclaimer and information on data protection.](#)



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