

IAP - IJPP CME 2023**DIETARY MANAGEMENT IN INBORN ERRORS OF METABOLISM*****Umamaheswari Balakrishnan**

Abstract: *Inborn errors of metabolism are individually rare, but collectively common. They pose challenges not only in diagnosis, but also in management. Dietetic interventions/ therapy plays a key role in both acute and chronic management of inborn errors of metabolism and helps prevent long-term complications. Adequate knowledge of diet therapy helps the clinicians effectively manage these children. This review article elaborates on the dietary principles and diet management in aminoacidopathy, organic acidemia, urea cycle disorders, fatty acid oxidation disorders, galactosemia, glycogen storage disorders and gluconeogenetic defects. The role of breast milk feeds in these disorders and the dos' and don'ts in diet therapy are also discussed.*

Keywords: *Inborn errors of metabolism, Diet, Dietary management.*

Points to Remember

- *Dietary management is the mainstay of therapy for disorders of intermediary metabolism.*
- *Reduction in substrate and providing deficient metabolite are the main principles of management which are based on diet therapy.*
- *Breast milk is the main source of natural protein during infancy and should not be stopped with exceptions being infantile onset VLCADD and galactosemia.*
- *Alteration in diet should be based on biochemical and clinical findings.*
- *A multidisciplinary approach is recommended in the management of IEM.*

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