

## 6. Outcome of Organic Acidemia in Indian Children

**Dr. Anil B. Jalan**

---

**Introduction:** - With a population over 1 billion and annual birth of about 28 million babies, the number of children born with Inborn errors of Metabolism is expected to be quite high. In absence of any formal newborn screening it is impossible to even guess the number of actual babies born alive with any IEM, especially Organic Acidemias. As regards organic acidemia, M.Muranjan et al has shown an incidence of 32 out of 231 children (13 %) with organic acidemia and overall mortality of 9.3 %. Out of 1,844 children investigated for IEM in last 7 yrs, we had 88 children with Organic Acidemia (4.77%) and a mortality of 37.5 % (33/88). Amongst the 6 commonest Organic Acidemias we had MMA 13 (14.77%), Propionic Acidemia 11(12.5 %), GA Type I -10(11.36 %), FDPase def. 9(10.23 %), Biotinidase deficiency 8(9.09 %) and MSUD 7(7.96 %). Mortality rates were - MSUD (100%), GA Type II (100 %), HMG CoA Lyase def. (100 %) and Propionic Acidemia (55%). Neurological affection was 100 % in GA Type I and quite high in MMA (53.85 %).

**Conclusion:** - Organic acidemias are common in Indian children. Unfortunately they are detected late, only after the symptoms have appeared. Most of the times, the diagnostic facilities are not available immediately and the cost of diet and medicines is beyond reach of most of the Indian patients. In such a situation the mortality is unacceptably high and those who survive have high rate of neurological affection and other complications.