where health is a State subject, difficulties arise in ensuring strict quality control in large number of small blood banks and its failure to take punitive measures for those who fail to follow its guidelines. Other feasible option may be that all small blood banks may be closed and blood banks should be attached with major medical institutions or organizations who are willing for multiple strict internal/external quality control for safety of blood and blood products. Thus, these blood banks will have sufficient blood for its optimal use by providing varius blood components. In addition, plasma fractionation unit can be part of at least few major blood banks for preparation of coagulation factors.

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Maple Syrup Urine Disease-2-4 DPNH Test as a Routine in Highly Sick Newborns

Only a few cases of maple syrup urine disease (MSUD), a common disorder of aminoacid metabolism have been reported in Indian literature(l,2). We report a case here to emphasise the utility of 2-4 DPNH test in seriouaIy ill new borns for early diagnosis of MSUD, in order to prevent neurodevelopmental sequalae of diseases resulting from accumulation of metabolites. MSUD results from a defect in the metabolism of leucine, valine and isoleucine(3). The accumulation of branched chain aminoacids leads to neurotoxicity and these metabolites are excreted in urine and sweat to produce the typical smell of maple syrup.

A male baby born to a third gravida mother of non-consanguineous marriage, was normal for the first five days. He subsequently developed excessive crying with refusal to feed. On the 8th day, he developed generalized tonic, clonic seizures. Pertinent investigation such as septicaemic screen, ultrasound and CT scan of brain were normal. The child did not respond to any therapy. On 13th day, abnormal smell was detected from body and urine. Blood ammonia, glucose, urea, pH, and fresh urine examinations were all normal. However, ketone bodies were positive in urine with positive ferric chloride and 2-4 DPNH test.

Aminoacidogram showed increased level of leucine, isoleucine and valine. The diagnosis was confirmed as MSUD by enzyme assay in leukocytes and fibroblast culture in New York University. On follow up, the child showed developmental delay with normal physical growth, which might have occured becaused of delay in diagnosis resulting in damage to neonatal brain by the accumalated branched chain aminoacids and their derivatives. On retrospective evaluation, there was a family history of unexplained deaths of two babies in early infancy following some respiratory problems.

This case demonstrates the typical way in which MSUD presents in infancy. MSUD should be suspected alongwith other causes in any severely sick new born. 2-4 DPNH test can be undertake in urine in any suspected new born before the typical smell develops. This will be helpful to make any early diagnosis. If the disease is diagnosed early and diet therapy is started accordingly, delay in mental development can be avoided(4). MSUD children need special diet which unfortunately is not available very easily in India. Because of the high morbidity and mortality associated with the disease, prognosis of the disease remains guarded.

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