Congenital Chylous Ascites and Lymphedema in Down's Syndrome

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Sir,

Chylous ascites is extravasation of chyle into the peritoneal cavity and a rare clinical entity in neonatal period. Even though Congenital chylous ascites and lymphedema has been associated with chromosomal disorders (Noonan syndrome, Turner syndrome and Klippel- Trnaunay-Weber syndrome), association with Down's syndrome has not been documented in the literature. Detailed PUBMED search revealed only one case with transient fetal ascitis in Down's syndrome [1]. However, Congenital chylous ascitis and lymphedema have not been reported yet in Downs syndrome. We report a case of chylous ascites with lymphedema in Down's syndrome.

A 21-d-old girl, born at 36 wk by normal delivery (Birth weight-2,300 g), presented from 2 wk of age with bilateral non-pitting pedal edema and ascites. (Abdominal circumference-45 cm). There was uneventful prenatal, natal and postnatal life. Mongoloid slant of eyes, brachycephaly, low set ear, long philtrum, simian crease, clinodactyly suggested Down's phenotype. Karyotyping confirmed the trisomy 21(47XX+21). On investigations, blood count and liver function test was normal except low serum albumin (2.3 g/dl). Urine examination, renal function test and skiagram of chest were within normal limits. Abdominal sonography and computerized tomography revealed no abnormality except ascitis. Abdominal paracentesis was done which showed milky white fluid, rich in triglycerides 5.9 mol/L, protein 1.3 g/dl and lymphocytes (430 cells/ μ L, lymphocytes -70 %). The baby

was treated conservatively, fed with skimmed milk, rich in medium chain triglycerides and low in long-chain triglycerides (LCT) and enhanced in protein content. Abdominal paracentesis was done once a week. With this treatment abdominal girth decreased from 45 cm to 36 cm within 3 wk. Then the baby was discharged and there was no recurrence of lymphedema or ascites up to 6 mo of follow-up.

Incidence of chylous ascites in hospital admission ranges from 1 in 1,50,000 to 1,75,000. Pathogenesis is understood poorly. Weber et al. [2] reviewed 51 congenital chylous ascites-30 children had no specific cause and 21 children had abnormalities of lymphatic channels. However, in infants, structural abnormalities of the lymphatic system are rare [3]. Paracentesis is the most useful diagnostic step. Microscopic examination of ascitic fluid demonstrates fat globules and predominance of lymphocytes with elevated triglycerides and protein. The treatment of congenital chylous ascites is conservative, including enteral feeding with a formula high in medium-chain triglycerides (MCT), low in long-chain triglycerides (LCT), enhanced protein content and repeated abdominal paracentesis [4]. If conservative approach fails, surgical intervention is recommended.

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