

If the Iris is Absent?

A 3-month-old boy born of a non-consanguineous marriage, was referred for nystagmus and a parietal bone defect. Physical examination showed horizontal nystagmus, macrocephaly, bulging anterior fontanelle, wide defect in the parietal bone, persistent neonatal reflexes and scissoring of the legs. The cornea was normal in shape and size, and showed a thin rim of iris bilaterally. The pupils were large and the foveal reflex was dull. Motor DQ was 4-6 weeks. Severe spasticity, adductor spasm and a positive heel tap was noted. The presence of a normal patella was confirmed radiographically. Abdominal ultrasonography showed normal kidneys. Karyotype of the peripheral blood leukocytes showed 46 XY pattern with deletion of the segment p13 to p15 of the chromosome 11 in all the metaphases studied (46XY, del 11 p13 to p15). Parents refused a cytogenetic analysis. CT scan of the brain revealed defect in the left parietal bone with normal dura underneath. MRI of the brain demonstrated prominent fronto-temporo-parietal cortical sulcal spaces over the cerebral convexities without any evidence of mass effect or midline shift and marked cerebral atrophy. Our patient had bilateral aniridia and mental retardation with normal kidneys on ultrasonography. The parietal bone defect was a coincidental finding.

Congenital aniridia can be sporadic or inherited as an autosomal dominant or autosomal recessive trait(1,2). Majority of the cases of aniridia with deletion 11p13 are sporadic. Moore *et al.*(3) have suggested that single chromosomal

breaks are associated with isolated aniridia, while deletions of 11p13 result in all the components of WAGR Syndrome (Wilms' tumor, aniridia, ambiguous genitalia, mental retardation)(3). Sixty five cases have been documented in the world literature(2). This is probably the first case report from India.

Management includes ophthalmic care for the secondary complications of aniridia and early detection and treatment of Wilms' tumor. Serial follow up scans would be important to detect Wilm's tumor at an early stage, as approximately 68% of the patients with bilateral aniridia develop Wilms' tumor by 3 years of age(2). Prognosis largely depends on the degree of mental retardation and the occurrence of gonadoblastoma or nephroblastoma. Life span and intelligence are normal in cases of isolated aniridia. Parents need to be subjected to cytogenetic analysis to rule out a balanced translocation. Prenatal diagnosis can be offered by doing a cordocentesis and a karyotype in cases where abnormality is detected in parents.

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Suthi Feeding: An Experience

Man discovered bottle feeding as an alternative method, in situations where breastfeeding cannot be given. With increase in use of "bottle", more and more hazards of bottle feeding were recognized. The campaign against bottle feeding was started. A concept of "Baby Friendly Hospital Initiative" has grown to promote, protect and support breastfeeding. However, a suitable alternative method of feeding in cases where expressed breastmilk is given or breastfeeding cannot be given at all, is still in question. Feeding with "spoon and bowl" (Katori-chammach) was considered to be a reasonable alternative as better cleanliness can be achieved.

In Nursery setting, particularly in our circumstances, where sufficient staff is lacking, it appears to be impractical to some extent to feed every neonate with "spoon and bowl", considering the time it consumes. For this reason other utensils were also evaluated and "Suthi" (Fig. 1) was considered a better alternative.

Suthi is a traditional "Sindhi" utensil

made of silver and presented by maternal grandparents to the child for drinking water. This vessel made up of stainless steel is also available and costs Rs. 7 to 10. It is a small semicircular vessel measuring 10 or 20 cc with a long semicircular beak having curved rounded margins. Salient features and advantages of Suthi feeding are: (i) It is a shallow vessel with a broad upper surface, so that it can be cleaned thoroughly and easily, thus preventing all the hazards of a deep vessel like bottle; (ii) Its narrow beak can be put directly into the mouth of neonate especially prematures and low birth weight babies with relative



Fig. 1. Photograph of Suthi