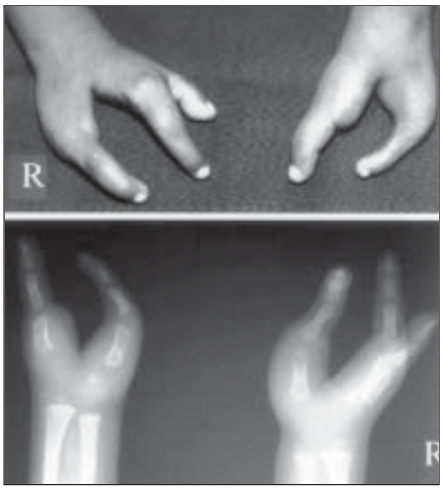

Figure 1: Photograph and X-ray of the hands. X-ray shows agenesis of two metacarpal bones on the right side and three on the left side with phalanges attached to them, resulting in a characteristic split hand malformation

Split-hand/split-foot malformation associated with maternal valproate consumption

Sir,

We examined a 3.5-year-old boy with congenital malformation of both upper limbs. His mother was taking sodium valproate 800 mg daily for complex partial seizures throughout pregnancy. There was no history of antenatal infection or exposure to any other medications, alcohol, smoking, or exposure to X-rays.

Both hands of the boy were malformed [Figure 1]. Below the wrist, his palms were split into two parts, the distal end of which had two fingers on the left and three fingers on the right side. The fingers and parts of the palms could be moved volitionally. He had mild hypertelorism and mild equinovarus deformity of the right foot. Rest of the examination was normal.

X-rays [Figure 1] showed agenesis of two metacarpal bones on the right side and three on the left side with phalanges attached to them, resulting in a characteristic split-hand malformation. There was only one carpal bone on the left side and none on the right side. A detailed pedigree evaluation for three generations failed to reveal any congenital malformations or consanguinity.

Split-hand/split-foot malformation (SHFM) is a limb malformation involving the central rays of the autopod, presenting with syndactyly, median clefts in hand or foot and aplasia or hypoplasia of metacarpals or metatarsals. Failure to maintain median apical ectodermal ridge (AER) signaling is the main pathogenic mechanism for which genetic causes are implicated. Five loci for SHFM have been mapped: SHFM1 on chromosome 7q21, SHFM2 on chromosome Xq26, SHFM3 on chromosome 10q24, SHFM4 that is caused by mutation in the TP63 gene on chromosome 3q27, and SHFM5 on 2q31. SHFM may occur as an isolated entity or as part of a syndrome. Scherer *et al*. in 1994[1] have classified ectrodactyly in to nine types after reviewing the published literature on clinical and genetic data. Both dominant and recessive pattern of inheritance have been documented in SHFM.

Our proband had no family history on pedigree evaluation for three generations and he was born of a nonconsanguineous parentage. In the reported case, prenatal exposure to valproate appears to be the only risk factor involved, but a chance association cannot be excluded.

Several limb reduction deformities have been reported with the use of valproic acid in clinical[2]–[7] and experimental settings.[8] But split-hand malformation deformity in relation to valproate exposure has not been reported earlier.

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Accepted on 08-9-2004