Letters to the Editor

Familial Radial Dyplasia with Renal Ectopia

Radial dysplasia is a congenital deficiency of structures on the radial border of the arm varying from mild shortening to complete aplasia of the radius along with proportional deficiency of muscles, nerves and vessels(1), frequently (70%) associated with non-limb abnormalities(2). The dysplasia is often seen as part of a more generalized syndrome, for example, Holt-Oram syndrome, Fanconi's syndrome, TAR syndrome, trisomy 17 and VATER association. We report radial dysplasia in three generations of a family and in two generations it was associated with crossed fused renal ectopia without any other systemic abnormality.

A four-day-old full term male baby normally delivered to a 35-year-old 3rd gravida mother presented to us. The baby had bilateral radial club hands with different degrees of radial hypoplasia on the two sides. Left forearm was shorter than the right one and left thumb was not well developed. Anthropometric parameters were within normal limits. The palate was high arched and the neck was short. There was no other congenital anomaly detectable on clinical examination. There was no facial dysmorphism.

X-ray of both upper limbs showed hypoplasia of radius, more on the right side. Moreover, the first metacarpal was also absent on the left side. Remaining skeletal survey did not reveal any abnormality except for bilateral cervical ribs. Abdominal ultrasongraphy revealed right crossed ectopic kidney that was malrotated and fused to the lower pole of left kidney. Intravenous pyelography revealed hydronephrotic changes of ectopic right kidney. Hemogram, renal functions, serum electrolytes, EKG, and echocardiography were normal. The baby was diagnosed as a case of bilateral radial club hands with right crossed fused renal ectopia and hydro-nephrosis.

Previously two children were born with no obvious anomaly or dysmorphism. The first child, conceived after 11 years of marriage, was a stillborn male; the second child, also a male had died at the age of six months' after a brief febrile illness.

The child's maternal grandfather as well as mother has club hands. Mother had bilateral radial club hands with absent thumb on left side and rudimentary thumb on right side. Maternal *X*-rays of hands showed bilateral absent radii and short ulna. The first metacarpal was absent bilaterally, there were two phalanges attached to the proximal phalanx of index finger on the right side. The abdominal ultrasound of the mother also revealed right crossed fused ectopic kidney. Grandfather was not available for work-up. There was no history of malformations on paternal side.

Factors such as intrauterine compression, an inflammatory process, vascular insult, maternal drug exposure (thalidomide, insulin), and irradiation have all been raised as possible etiologic causes. The pattern of inheritance may be autosomal dominant or recessive. In the present case the inheritance seems to be autosomal dominant. There may be associated chromosomal abnormalities, including trisomy 13, 18 and 21. Although chromosomal studies could not be carried out in our patient due to financial constraints, the pattern of

inheritance and co-existence of renal ectopia with radial dysplasia suggests some malformation syndrome with autosomal dominant inheritance.

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Plasmapheresis in Acute Disseminated Encephalomyelitis

I read with interest the recent report of plasmapheresis in childhood acute disseminated encephalomyelitis (ADEM) resulting in remarkable recovery(1). However, I would like to make certain observations.

Though the indication mentioned for resorting to plasmapheresis in the report is the unaffordability of intravenous immunoglobulin (IVIG) therapy, I wish to highlight other important reasons for preferring plasmapheresis over IVIG. Firstly, IVIG is known to exert its immunomodulatory effects for a prolonged period of time. The mean duration of action of IVIG is 53 days and the half life of immunoglobulin in the serum is 3-4 weeks(2). Therefore, employing plasmapheresis immediately after IVIG therapy would result in removal of circulating IVIG, thereby giving little time for IVIG to show its efficacy. This removal could be avoided by opting for IVIG therapy only after plasmapheresis has failed. Secondly, there are published reports of plasmapheresis succeeding even in IVIG-refractory cases of ADEM(3,4).

The probable reasons, IVIG is preferred over plasmapheresis in ADEM, are its ease of administration, the lack of plasmapheresis facilities in many centers and a fear of treatment-related complications with plasmapheresis. However, low-volume (manual) plasma exchanges have been shown to be efficacious in ADEM and can be performed with little training even in smaller centers(5). At the same time, low-volume exchanges are relatively safe too. However, I agree with the authors that randomized controlled trials are required to decide the most effective volume of plasmapheresis required in ADEM.

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