LETTER TO EDITOR (VIEWERS CHOICE)

DOWN SYNDROME AND HEMIHYPERTROPHY

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A 9 months old Hindu male child from Midnapore district of West Bengal having Down syndrome born of non consanguineous marriage, after three successive pregnancy losses, with maternal age 33 years presented with asymmetric enlargement of the right half of his body from birth. The baby had a history of delayed cry after birth. He weighed 5.6 kg, length was 69 cm and head circumference was 45 cm. On physical examination, there was clear asymmetry between his right and left upper and lower limbs. Right midarm circumference was 12.5 cm, left was 10.5cm, the circumference of the right leg at the level of mid thigh was 20cm, while the left measured 17.5cm. There was considerable leg-length discrepancy with the right leg being 2cm longer than the left. Examination of the chest and abdomen were unremarkable.

Hemihypertrophy is best defined as asymmetry between right and left sides of the body to a greater degree than can be attributed to normal variations (1). Patients with hemihypertrophy have increased propensity to develop malignant embryonal tumors like Wilm;s tumor, hepatoblastoma and many others. (2,3). Hemihypertrophy can be classified as congenital or acquired. Congenital hemihypertrophy may also be classified as total or limited. Total hemihypertrophy involves all organ systems, including ipsilateral paired organs, whereas those who have limited hemihypertrophy have only muscular, vascular, skeletal

or neurological involvement. Limited forms include classic hemihypertrophy (ipsilateral involvement of upper and lower limbs) or segmental (limited to an upper or lower limb), facial or crossed (involving contralateral upper and lower limbs) (4). A threshold of 5% difference is used to define abnormal asymmetry as suggested by Pappas & Nehme (5) and Andersen (6) et al. Thus discrepancy between lengths of limbs is 1.3 cm at 1 year of age, 2.3 cm at 5 years of age, 3.2 cm at 10 years of age and 4.1 cm at 18 years of age. Our case had congenital hemihypertrophy, limited form, and classic type with a limb-length discrepancy of 2cm, and a limb circumferential disparity of 2 to 3 cm. Several conditions have been found to be associated with hemihypertrophy such as Beckwith-Weideman Syndrome, Neurofibromatosis, Proteus syndrome and Klippel Trenaunay Weber Syndrome (7-11). Hemihypertrophy with Down syndrome has not been reported earlier.

As there is risk for malignancy, our patient is being followed up at regular intervals with serial ultrasound of abdomen.

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