LETTER TO EDITOR (VIEWERS CHOICE)

DOWN SYNDROME AND HEMIHYPERTROPHY

Kaustav Nayek, A Banerjee, N Ghosh, S Pati

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 10.5 cm, the circumference of the right leg at the level of mid thigh was 20 cm, while the left measured 17.5 cm. There was considerable leg-length discrepancy with the right leg being 2 cm 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 Wilms 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

REFERENCES

2. Allington NJ, Kumar SJ, Guille JT. Clubfeet associated with amniotic band formation and disruption of foetal parts due to compression or ischemia from amniotic bands. (fibrous cords) form which strangle surface of mesoderm. (6) A decidual reaction occurs comes in contact with sticky mesoderm on chorionic amnion without rupture of chorion causes transient oligohydramnios due to loss of amniotic fluid through amniotic to extra-embryonic coelom through defect and initial permeable chorion. The foetus passes from limb 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 Wilms 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).

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

REFERENCES
5. Pappas AM, Nehme AM. Leg length discrepancy associated with hypertrophy. Clin Orthop Relat Res. 1979; 144: 198-211

From: Department of Pediatrics, Burdwan Medical College and Hospital, West Bengal, India.

Address for Correspondence: Dr Kaustav Nayek, Laxmipurmath, Kantapukur, Burdwan-1, West Bengal. Email: kaustav25@yahoo.co.in

E-published: 1st February 2012 Art#9

DOI No. 10.7199/ped.oncall.2012.9