Poland Syndrome with Dextrocardia

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The incidence of Poland syndrome is estimated as 1: 25,000 births, the male to female ratio being 3:1(1). This sporadic condition is characterized by unilateral absence of one or more portions of the pectoralis major (usually sternal head) together with ipsilateral hand abnormalities in form of syndactyly and/or hypoplasia of hand. In 25-30% of cases, ipsilateral rib or costal cartilage defects or absence of breast or nipple may occur(1,2). Association of dextrocardia with Poland Syndrome is extremely rare(2). Till data only two cases of Poland Syndrome with dextrocardia have been reported in the World Literature. We report the third case of isolated Poland's anomaly with dextrocardia.

Case Report

A male term infant, weighing 3520 g, products of an uncomplicated 38 weeks pregnancy and delivery was born with an Apgar score of 9, 10, 10 at 1, 5 and 10 minutes. The parents were unrelated, young and healthy at the time of conception. There was no history of maternal irradiation or drug intake. On admission to Nursery, the congenital abnormalities observed were left sided depressed hemithorax with loss of normal chest contour, areolar hypoplasia, widened III intercostal space close to parasternal area and ipsilateral symbrachydactyly of index and middle finger. Systemic examination revealed dextrocardia, liver 1.5 cm below right costal margin. Radiographs showed dextrocardia (*Fig. 1*) without abdominal heterotaxia with III rib hypoplasia with hypoplasia of Phalanx of Index and middle finger of left hand with cutaneous fusion. ECG showed upright "P" wave in aVR and inverted "P" in lead I suggestive of dextrocardia. Echocardiography confirmed dextrocardia without inversion or valvular, ductal or septal abnormality. Abdominal ultrasound confirmed nor-

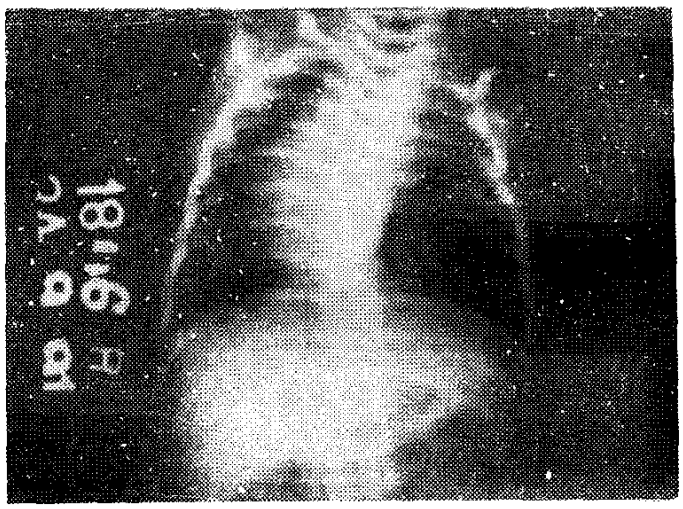

Fig. 1. Radiograph showing Dextrocardia

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mal right side liver and normal kidneys on both sides. Hemogram and peripheral smear did not reveal any abnormality.

Discussion

Alfred Poland in 1841 described the association of aplasia/hypoplasia of the sternal head of the pectoralis major muscle with ipsilateral symbrachydactyly(2,4), Since then several cases of Poland Syndrome have been reported with varying associations like ipsilateral Moebius syndrome(4,5), absent kidney and upper limb musculature(2,4), rib deformities, Sprengel's deformity, hemivertebrae and radio-ulnar synostosis and axillary webs(6,7).

Dextrocardia has been reported only with two cases of isolated Poland Syndrome, one had associated acute lymphatic leukemia(2) and other had absent incisors(2). Dextrocardia in association with Poland and Moebitis syndrome has been reported twice(2). Our patient was isolated case of Poland's anomaly with dextrocardia without any evidence of leukemia at the time of diagnosis.

Embryogenesis of this anomaly is unknown but it has been suggested that primary defect may be in the development of the proximal subclavian artery with early deficit of blood flow to the distal limb and pectoral region(1,8,9).

Surgical management of Poland Anomaly in females includes latissimus dorsi augmentation to anterior chest with silicone gel prosthesis placement in submuscular position to augment breast movement. In males transfer of latissimus dorsi is done if abduction of involved shoulder is severely affected(1,3).

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Celiac Disease in Insulin Dependent Diabetes Mellitus

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Diarrhea and failure to thrive in a patient with diabetes mellitus may often be

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