- Introduction -

Congenital short femur is an uncommon congenital deformity chararacterized by defective morphogenesis (dysgenesis) of the proximal portion of the femur (Aitken, 1969).

The condition may be unilateral or bilateral and is often associated with other congenital anomalies. The incidence of this complex defect is 1 in 50.000 (Grill, Dungl, Steinwender and Hosny, 1993).

The manifestations of femoral dysplasia range from an intact femur approximately 60 percent of the length of normal leg to a subtotal absence of the femur in which only the femoral condyles remain, often with a congenital fusion of the knee joint. Two groups were defined: group(I) consisted of those with congenital hypoplastic femur in which the hip and knee could be made functional and where - in some positions at least-leg equalization was possible; group (II) consisted of those with true proximal femoral focal deficiency where the hip joint was never normal and the knee joint was always useless (Gillespie & Torode, 1983).

There are four major biomechanical losses in children with lowerextremity skeletal deficiencies:-

- (1) inequality of limb length,
- (2) malrotation,
- (3) inadequacy of the proximal musculature, and
- (4) instability of the proximal joints (Epps, 1983).

The established classifications of congenital abnormalities of the femur concentrate on detailed examination of radiographs (Gillespie & Torode, 1983).

The percentage of inhibition in the involved femur compared with the contralateral side was found to be constant in unilateral cases; in bilateral cases relative growth was also demonstrated to be constant. A femur was considered normal if it had a normal roentgenographic appearance and was within one standard deviation from the mean for skeletal age (Koman et al., 1982).

Simple hypoplasia of the femur may possibly have a multifactorial genetic background since it is associated with other minor abnormalities of the limbs in these families, whereas environmental factors only are associated with the more severe femoral defects (Hamanishi, 1980).

Proximal femoral focal deficiency may be confused with other congenital femoral deformities, such as coxa vara and generalized dysplasias. Coxa vara with femoral bowing is associated with varus of the femoral neck, unlike PFFD, in which the varus is in the subtrochanteric region (Grogan et al., 1987).

The major problems facing these children with such deformities are due to limb length inequality. The goals of treatment are to anticipate the ultimate discripancy, to maintain normal function of knee and hip joint, to deal effectively with the associated anomalies, and to minimize the number of operations and hospitalizations (Grill, Dungl, Steinwender, and Hosny, 1993).