**Case Reports**

Poland Syndrome with Unusual Associated Anomalies: A Case Report

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**Introduction**

Poland syndrome is marked by unilateral absence or hypoplasia of pectoralis major muscle (most frequently involving the sternocostal portion) and a varying degree of ipsilateral hand anomalies. It has been estimated that 10% of patients with syndactyly of the hand have the Poland sequence. In 1841, it was first reported by Alfred Poland¹. Absence or hypoplasia of the breast and nipple have also been reported in this. Other associations observed are abnormal rib, dextrocardia, only in left sided muscle hypoplasia, oligodactyly, preaxial polydactyly of upper extremity, absent radius/ulna/humerus, scapula anomaly, microcephaly, encephalocele, abnormality in GIT and biliary tree, ureteric anomaly and vertebral segmentation anomaly². The incidence is one in 7000 to one in 100,000 live births³. The vast majority of cases are sporadic⁴ but familial cases have been reported, compatible with an autosomal dominant mode of inheritance. Males are three times more affected than females and is 75% right sided⁵,⁶. We report a case of Poland syndrome in a newborn with the involvement of left side and ipsilateral preaxial polydactyly of foot with bifid hallux and synostosis of 1st metatarsal bone instead of the documented syndactyly/polydactyly/sympbrachydactyly of phalanges of the hand.

**Case Report**

A male, inborn newborn, only issue of a nonconsanguinous parents was brought to the neonatal care unit of Bangabandhu Sheikh Mujib Medical University 2 hours after birth because of respiratory distress with grunting. Baby cried immediately after birth but developed grunting within half an hour. On examination, birth weight was 2100 gm, length was 44cm which fell on 25th centile, OFC was 33 cm which fell on 75th centile, face was normal, anterior chest wall was depressed on the left side and the breast nipple was hypoplastic in comparison to right one (Fig.-1).

![Fig.-1: Showing depressed chest wall, hypoplasia of left pectoral muscle with hypoplastic nipple](image_url)

Respiratory rate was 74/min, heart rate was 140/min and all the heart sounds were audible on the right side of the chest. Upper arm, forearm including hand on both sides were normal. Preaxial polydactyly with bifid hallux was evident on the left foot (Fig.-2). No wasting was observed on the gluteal region. Other systems were clinically normal. He was diagnosed as a case of preterm (35 weeker) AGA, LBW with RDS and multiple congenital anomaly.

His haemoglobin and complete blood count was normal. Capillary blood glucose, serum electrolyte and calcium level were also within normal range. On third day, serum bilirubin was estimated to be 230 micromol/L. CRP of blood was 48mg/L i.e, above
normal level (6 mg/L), indicating infection. X-ray chest revealed dextrocardia and hypoplastic ribs on the left side (Fig.-3).

Ultrasonographic scan confirmed the presence of hypoplasia of left sided pectoralis major muscle. Doppler echocardiography showed mild patent foramen ovale, tricuspid regurgitation and pulmonary hypertension. X-ray of left foot revealed synostosis of 1st metatarsal bone (Fig.-4).

He was managed with O₂ inhalation, IV fluid and antibiotic. Later on he received phototherapy.

Discussion

Mc Gillivary and Lowrys ascertained 44 cases of Poland syndrome in British Columbia. There was variable involvement of the hands and arms with 4 patients having normal hands. Riccardi described unilateral gluteal hypoplasia and brachysyndactyly and proposed that it was the lower limb equivalent of the Poland anomaly. In support of Riccardi's view, Parano et al reported on an extended family in which 3 individuals, all women had unilateral gluteal hypoplasia and the propositus in addition had unilateral hypoplasia of a pectoral muscle. Another relative in this family had unilateral symbrachydactyly of the distal phalanges of one foot. Corona - Rivera et al reported another instance of a possible lower extremity counterpart of Poland sequence. They proposed a vascular origin which would involve the external iliac artery supply analogous to the disruption of the subclavian artery supply in the upper extremity Poland sequence. Nanjeeba reported a case of Poland sequence in a girl with congenital haemangioma and polydactyly in hands of the right side but no syndactyly, brachydactyly or oligodactyly. In our case in addition to hypoplasia of left sided pectoralis major muscle, nipple hypoplasia, dextrocardia and hypoplastic ribs, preaxial polydactyly and bifid hallux was evident on the left foot though no wasting of the gluteal region of the same side was obvious clinically. Radiologically, synostosis of metatarsal bone was
seen which contrasts the traditional finding of synostosis of phalanges leading into symbrachydactyly. Bifid left hallux with syndactyly was previously reported by Kabra M12.

Conclusion
Poland syndrome is commonly described as hypoplasia of pectoralis major muscle and syndactyly of hand. But preaxial polydactyly with bifid left hallux and synostosis of metatarsal bone in addition to dextrocardia and mild congenital heart disease may be included as one of the components of this syndrome.

References
2. WWW.orpha.net /static/GB/ Poland anomaly Update. 10.12.2006.