



## Poland Syndrome: A Case Report

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### ABSTRACT

Poland syndrome is a rare entity with average incidence of one in 30000. Named after sir Alfred Poland, Poland syndrome includes the features of under development of the chest muscle on one side of the body, cutaneous syndactyly on the same side of the body and some time the lack of development of the breast and bone. We report a case of 1 year female child presenting with the features of Poland syndrome

### KEYWORDS

Poland syndrome, cutaneous syndactyly, athelia, radioulnar synostosis

### INTRODUCTION:

The Poland's anomaly was first described in 1841 by Sir Alfred Poland as a syndrome

presenting with absence or underdevelopment of pectoralis major muscle, associated in some cases with a hypoplasia of the breast, an agenesis of 2,3,4 and 5 ipsilateral costal cartilage, an athelia, and an ipsilateral webbing of the fingers (cutaneous syndactyly)[1,2]. Therefore the Poland syndrome may occur with different gravity.

Currently, it is assumed that Poland syndrome is characterized by a missing sternocostal bundle of the pectoralis major muscle [3]. Its incidence is difficult to determine, but current estimates range between 1:7,000 and 1: 100,000 births, with higher frequency among males (ratio, 2:1 – 3:1). In 75% of the cases, it is located on the right hemithorax in the unilateral form[1,3,4,5,6].

Affected individuals may have variable associated features, such as underdevelopment or absence of one nipple including the areola and/or patchy absence of hair in the axilla[1,4].

**In females, there may be underdevelopment or aplasia of one breast and underlying (subcutaneous) tissues[7].**

In some cases, associated skeletal abnormalities may also be present, such as underdevelopment or absence of upper ribs; elevation of the shoulder blade (Sprengel deformity); and/or shortening of the arm, with underdevelopment of the forearm bones i.e., ulna and radius [1,4]

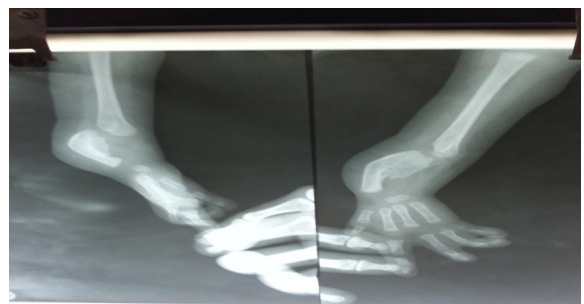
### CASE REPORT:

1 year female child was admitted with complains of fever, cough and cold. On examination this girl child had cutaneous syndactyly and absence of nipple (athelia) and chest wall deformities on left side of the body. (figure 1)



**Figure 1 : athelia ,chest wall deformity and syndactyly on left side of the body**

**X ray left forearm showed radial hypoplasia with radioulnar synostosis along with soft tissue fusion of 4<sup>th</sup> and 5<sup>th</sup> fingers.{fig2}**



**Fig 2 : radial hypoplasia with radioulnar synostosis**

**Chest xray reveals fusion of ribs as shown in figure 2.in this child 1<sup>st</sup> 2<sup>nd</sup> 3<sup>rd</sup> 4<sup>th</sup> ribs are fused together and 5<sup>th</sup>, 6<sup>th</sup> ribs fused together and 7<sup>th</sup>, 8<sup>th</sup> ribs are fused together.(fig 3)**



**Fig 3: chest x ray showing fused ribs**

#### DISCUSSION:

Poland syndrome, as first described by Alfred Poland in a 26-year-old male, most often occurs sporadically. The exact etiology of the Poland syndrome is unknown. It is assumed that the aplasia of the pectoralis muscles and associated chest defects, as the athelia, aplasia of costal cartilages, are consequences of an interruption of early embryonic blood supply of subclavicular artery branches [5,6]. A combination of the blockage of various branches could lead to different manifestations of the Poland syndrome.

It is known that thoracic wall is supplied by medial thoracic branches, intercostals artery, and the thoracic artery from axillary artery, the thoraco-acromial artery and the lateral thoracic artery. All these branches come from the subscapular artery or axillary artery.

The interruption of the blood supply is caused by thrombus or embolus, which prevent the blood to reach the developing tissue. Another cause of blood supply interruption is the misde-

velopment of vessels. However, there have been case reports of Poland syndrome associated with unusual defects, which can not be explained on the basis of compromised blood supply alone. On the other hand, Ferraro and colleagues [3] described an unusual presentation of the Poland's anomaly without any vascular alteration, raising the question as to the true pathogenesis of the Poland's syndrome.

Geneticists currently hold the view that Poland syndrome is rarely inherited and generally is a sporadic event. There are rare instances where more than one individual has been identified with Poland syndrome either in the immediate [6,9,10] or extended family [11,12,13]. Therefore, some authors believe that an inherited abnormal vasculature formation may be the central underlying mechanism for this condition.

Rare association between Poland syndrome with other organ system-related symptoms including microcephaly, cerebral atrophy, disorders in myelination, situs inversus or dextrocardia, hemivertebra, gastroschisis, paralysis of the cranial nerve or mental retardation, psychosocial retardation, hypospadias, and urinary system anomalies have also been reported.

Endocrine anomalies, melanosis, and an increase in the incidence of benign and malignant tumors may also be observed in Poland syndrome. Although the most commonly encountered malignant tumors are lymphoreticular tumours, such as leukemia and lymphoma, childhood solid tumours, such as neuroblastoma and Wilms' tumours, may also be observed [8].

Several reconstructive procedures are available to correct the functional and structural deformities associated with this syndrome. As for the chest deformity, customized silicone prosthesis is simply and safely used. Transposition of the latissimus dorsi muscle for soft-tissue reconstruction has been used by many authors with satisfactory esthetic and functional results [14].

#### Conclusion:

Poland syndrome has broad spectrum of skeletal, ocular, dental and maxillofacial anomalies. Multidisciplinary team approach is required for motor skills and co-ordination to gain better control over activities of daily living and patient counseling about this entity is very crucial.

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