
Case Report

Poland Syndrome: A Case Report

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Abstract

Poland Syndrome is a rare birth anomaly commonly found in males characterized by partial or complete absence of pectoralis major muscle on one side of the body (mostly right side), along with some other associated anomalies like asymmetry of upper limb, hypo-plastic nipple and syndactyly of the effected

side. A 4-month old male baby presented with complaint of fever and respiratory distress, was admitted and treated as case of bronchopneumonia. The patient was ultimately diagnosed as a case of Poland syndrome.

INTRODUCTION

Sir Alfred Poland described Poland syndrome as chest wall anomaly in the Guy's Hospital Gazette in 1841, while still a medical student based on findings of one cadaver dissection.¹ Poland syndrome is noted for the underdevelopment or absence of the chest (pectoralis) muscles on one side of the body as well as webbing of the fingers (cutaneous syndactyly) on the hand of the same side (ipsilateral side) of the body. Malformation of interior chest wall and breast; dextrocardia; lung herniation; renal, vertebral and lower limb malformation have been described as other usual anomalies in Poland syndrome.² Incidence of Poland Syndrome is difficult to determine but current estimates range between 1:7000 and 1:100000 births with high frequency among males. In 75 % of the cases, it has been located on the right hemithorax in the unilateral form.³

We present the first documented case from Faisalabad, Pakistan. This paper is aimed at bringing awareness to health professional about this rare congenital condition.

CASE REPORT

A 4-month old male, third born of consanguineous parents, developmentally normal, vaccinated according to EPI, presented with complaints of fever and respiratory distress for 1 day. There was no familial history of similar disorder. Examination revealed no facial dysmorphism, with weight of 4.5 kg, length 66 cm, Head Circumference 38 cm. The sternocostal part of pectoralis major was absent on left side, left

nipple was placed at lower level and there was prominence of costochondral junction of the affected side. There was asymmetry of left arm that was short with syndactyly of index and middle finger. Patient had pulse rate 124/min, respiratory rate 68/min, temperature 99°F, BP 80/60 mmHg, oxygen saturation at room air 94%. Apex beat was in 4th intercostals space in midclavicular line. Heart sounds were audible and there was bilateral equal air entry in both lungs with few crepitations. No other systemic abnormality was found.

Figure-1

Absence of left Pectoralis muscle and short left arm



Figure-2
Syndactyly of index and middle finger



Figure-3
X-ray chest showing normal heart and Ribs



Radiological examination of chest showed no abnormalities of ribs or heart. X-ray of affected limb was normal, ECG and abdominal ultrasound for renal pathology was also normal. Based on above mentioned physical anomalies, diagnosis of Poland syndrome was established. Patient was treated for bronchopneumonia and discharged after 4 days. No surgical treatment was offered. Family was counseled and patient was called for regular follow-up on outpatient basis.

DISCUSSION

The case of Poland syndrome we present is the first described in Faisalabad and is of pure presentation as it consists only on the unilateral aplasia of the pectoralis major muscle and syndactyly of the same side without

any other associated defects. However, it is presented in the left hemithorax while the right side is most frequently involved.³ The cause of Poland syndrome is uncertain. The disorder is currently considered "a nonspecific developmental field defect" occurring at about the sixth week of fetal development. According to one report, diminished blood flow through the subclavian artery that goes to the arm has been considered as a cause, but there is not enough proof for this idea. The exact cause of Poland anomaly is not known, but may result from the interruption of fetal growth at about the 46th day of pregnancy, when the fetal fingers and pectoralis muscle are developing.⁴

Poland's Syndrome occurs infrequently and does not run in families. The risk of reappearance of Poland's Syndrome in the family is very rare. For purposes of genetic counseling, Poland's Syndrome can be regarded as a random condition with an extremely low risk of being transmitted from parents to child.⁴

The surgical options for chest wall asymmetry depend on anatomical severity, gender, associated anomalies, and patient preferences. Treatment options include autologous fat injection, pedicled latissimus dorsi muscle transfer, transverse rectus abdominus musculocutaneous flaps, deep inferior epigastric artery perforator flaps, custom-made chest wall prosthesis, nipple-areola complex repositioning, mammary prosthesis, sternal/rib reconstruction, contralateral operations (to meliorate asymmetry), or a combination of these techniques.⁵ The understanding of the intervention options are complicated by the lack of detailed, long-term studies.

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