

Case Report

Poland Syndrome: A Case Study

MV Naveen Reddy^{1*} and M Rupa Reddy²

ABSTRACT

We report a rare case of Poland syndrome in a 22-year-old female patient. This is a syndrome defined by the unilateral absence or hypoplasia of the thoracic muscles with under development of one breast. A female patient presented with aesthetic concerns having hypoplasia of left breast and the nipple-areola complex small and displaced laterally. There were no other developmental problems with ribs, breast bone or upper extremity. The patient was surgically treated with breast reconstruction using a mammary prosthesis. The incidence of Poland syndrome ranges from 1 in 7000 to 1 in 1,00,000 [Fokin and Robicsek (2002). *Ann Thorac Surg* 2002;74(6):2218–25]. Ten percent of these patients also express some of the other features of the disorder. There is a male to female ratio of 2:1 to 3:1.

Keywords: Mammary hypoplasia, Aesthetic contouring, Mammary prosthesis, Nipple-areola complex, Sporadic, Pectoralis muscle

INTRODUCTION

Poland syndrome^[1] is a rarely recognised syndrome characterised by under development of one breast and the underlying pectoralis muscle. There may be an underlying developmental problem with the upper extremity. In 1841, Alfred Poland, a London medical student, reported some unusual anatomic findings after a cadaver dissection at Guy's Hospital^[2]. The subject of his report George Elt's left torso and upper extremity were underdeveloped. The Poland syndrome seems to be sporadic in nature (<1% risk of a second family member affected). Some patients have been linked to familial transmission, with unaffected parents giving rise to affected children. This mode of transmission is characteristic of a delayed dominant germ-line mutation.

CASE REPORT

A 22-year-old female patient reported to the gynaecology OPD with the underdeveloped left breast and was concerned with aesthetically unsatisfactory appearance

and future lactation function. O/E the patient had well-developed secondary sexual characters except hypoplasia of left breast with lateral deviation of nipple – areola complex. Other developmental problems of Poland's syndrome were absent. There was no other functional deformity. All the routine investigations were found normal. A routine x-ray chest showed normal chest bones. Mammography was suggestive of hypoplasia of left breast^[4].

The patient was surgically treated in view of her aesthetic concerns for breast contouring. She underwent breast reconstruction on the left side and a mammary prosthesis was placed to correct the asymmetry of the chest wall.

DISCUSSION

The precise mechanism causing the Poland syndrome has not been elucidated. Some data suggest disruption of the early embryonic blood supply in the subclavian or vertebral systems cause the syndrome during the 6th to 7th weeks of gestation^[3]. The cause of the disruption

^{1,2}Associate Professor, Department of OBG and Gynecology, Shadan Institute of Medical Sciences, Hyderabad, Telangana
*Corresponding author email id: mvnavin@yahoo.com

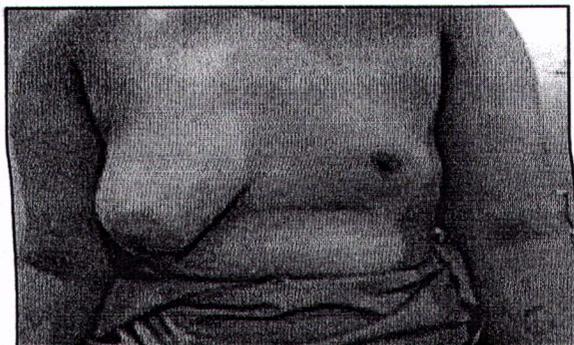


Figure 1: Poland syndrome: pre-operative photo



Figure-2: Poland syndrome: post-operative photo

has been postulated to be mechanical, focal compression of the vessels^[6]. However, alternative theories suggest that injury or developmental failure of the embryonic mesodermal plate during the 3rd and 4th weeks of gestation (which gives rise to the pectoralis muscle) can culminate in the Poland syndrome. Although highly variable, the Poland syndrome has several common features. The absence of the sternal head of the pectoralis major muscle is constant. The ipsilateral chest wall can suffer from several anomalies such as alopecia of the breast and axilla, lack of subcutaneous tissues, absent pectoralis minor muscle, absence or distortion of ribs two to five^[5]. Paradoxical motion of chest wall is observed in some. Lung herniation through the defect occurs in 8%. The latissimus dorsi, serratus anterior and external oblique muscle are occasionally absent. Ipsilaterallimb anomalies are present in 13–56% of patients, including brachysyndactyly as well as shortening of the limb.

The breast is affected in one-third of patients. Defects range from amastia to simple hypoplasia.

The nipple is often absent or elevated and hypoplastic. Patients, especially women, with the most mild form of Poland's syndrome often appear to be normal except for marked underdevelopment of a breast. The underlying pectoralis muscle is usually also underdeveloped but unrecognised. Associations have been drawn with renal abnormalities, hemivertebrae, leukemia, Mobius and Sprengel anomalies.

The severity of deformity has two anatomic variations. The simple (mild) form is the most common and the complex (severe) form. These differing types require different reconstructive solutions.

CONCLUSION

The Poland syndrome is a highly variable disorder of the thoracic musculature. The timing and technique of thoracic repair are based on the structures affected and the severity of the disorder^[7].

Instability of the chest wall should be corrected promptly. The optimal timing of the breast or aesthetic chest reconstruction is late in the second decade. The delay till the end of adolescence ensures that puberty has passed and the contra lateral chest and breast are fully developed, optimising the chances of obtaining symmetric results.

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MV Naveen Reddy and M Rupa Reddy

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