Scientific Research Publishing

Modern Plastic Surgery, 2024, 14, 99-107
https://www.scirp.org/journal/mps
ISSN Online: 2164-5280
ISSN Print: 2164-5213

Poland Syndrome: Breast Reconstruction Outcomes in 10 Years (2013-2023)

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How to cite this paper: Recio-España, F.I., Márquez-Espriella, C., Dávila-Díaz, R., Cuervo-Vergara, M.A., García, G.B., Campollo-López, P., Chama-Naranjo, A., Pérez-Benítez, O., Barlandas-Quintana, E., Garibaldi-Bernot, M. and Poblano-Olivares, E. (2024) Poland Syndrome: Breast Reconstruction Outcomes in 10 Years (2013-2023). *Modern Plastic Surgery*, **14**, 99-107.
https://doi.org/10.4236/mps.2024.144010

Received: August 28, 2024
Accepted: October 12, 2024
Published: October 15, 2024

Abstract

Summary: Poland syndrome is a rare congenital anomaly characterized by the partial or complete absence of the pectoralis major muscle and a wide spectrum of thoracic anomalies, predominantly on the ipsilateral side. These anomalies include hypoplasia or aplasia of the breast and its components, hypotrophy of subcutaneous fat, and absence of axillary hair, as well as hand deformities that can range from syndactyly to ectrodactyly. The aim of this study was to gather information about patients diagnosed with Poland syndrome at the Central South High Specialty Hospital of Petróleos Mexicanos over a period of 10 years and to identify their reconstructive algorithm. **Materials and Methods:** A retrospective, observational, and descriptive study was conducted to identify the population diagnosed with Poland syndrome at the "Central South High Specialty Hospital of Petróleos Mexicanos" during the period from 2013 to 2023. **Results:** The database of patients with Poland syndrome from 2013 to 2023 was analyzed, identifying a total of 8 patients with this diagnosis. Of these, 7 were women (90%) and 1 was a man (10%). The left side was more frequently affected (80%) compared to the right side (20%). The average reconstructive process required two surgical stages, mainly consisting of breast expander reconstruction (first stage) and replacement of the expander with an implant (second stage). **Conclusions:** Despite being a rare congenital condition, the volume of patients treated at the Central South High Specialty Hospital allows for improved diagnosis and contributes to their reconstructive process. The lack of diagnosis in the male population is notable, likely due to the absence of adequate screening.

Keywords

Poland Syndrome, Breast Reconstruction, Poland, Tissue Expander Breast Reconstruction

1. Introduction

1.1. Background

Poland Syndrome (PS) is a rare congenital pathology characterized by the unilateral absence of the sternocostal head of the pectoralis major muscle, along with ipsilateral chest wall and limb anomalies, with brachymesophalangia being the most common.

Although Lallemand, a French author, was the first to describe Poland Syndrome in 1826, the eponym "Poland Syndactyly" was coined by Clarkson, who published an article with the same title in 1962, honoring Alfred Poland based on research by Brown and McDowell [1] [2].

1.2. Epidemiology

The current incidence of Poland Syndrome is estimated to be between 1 in 30,000 and 1 in 32,000. It represents 14% of cases of breast aplasia and occurs in up to 10% of patients with syndactyly. There is a male predominance (M/F = 3:1), and the right side is involved in 60% - 75% of sporadic cases. In female patients, there is no significant side predominance [3].

1.3. Pathophysiology

This dysmorphia, sometimes referred to as Poland Sequence (PS), is primarily thought to result from an interruption of the blood supply to the thoracic limb primordium during the sixth week of gestation [4]. Dustagheer observed that this week coincides with the division of the pectoralis muscle into its clavicular and sternocostal portions, as well as the differentiation of the sixth aortic arch into two main branches: the subclavian and vertebral arteries. The formation of fingers and interdigital tissue apoptosis to create the interdigital spaces also occurs during this time. This disruption leads to hypoplasia of the subclavian artery or one of its branches, which determines the clinical severity of the anomaly. However, the pathophysiological evidence supporting this causality remains inconsistent [5].

1.4. Clinical Features

Poland Syndrome is characterized by a deficiency of the pectoralis major muscle and presents with a wide range of phenotypic variability, including partial agenesis or deformity of the rib cartilage, hypoplasia or aplasia of the breast and nipple-areola complex, axillary fold, and subcutaneous tissue, as well as sternal deformities and anomalies of the ipsilateral arm [6] (**Figure 1**).

The morphological classification of hand and upper limb malformations enhances the understanding of prognosis, function, and appearance in PS. The anatomical features of the thorax have led to the division of PS into two main variants: the simple or mild form, and the complex or severe form. Foucras has classified patients with PS according to thoracic deformity into three degrees based on severity (**Table 1**) [7] [8].

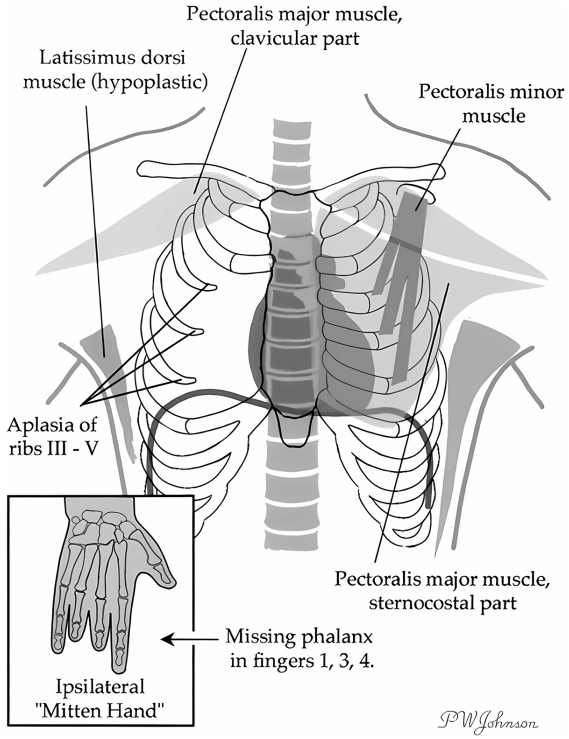

Figure 1. Anatomic representation of classic Poland's syndrome manifestations.

Table 1. PS Foucras Classification. Description, morphology, reconstruction 3D and CT (Translation of Pinsolle V *et al.*).

Grade I	Grade II	Grade III
Mild deformity	Moderate Deformity	Severe Deformity
Pectoralis major hypoplasia	Pronounced pectoralis major aplasia and other muscles hypoplasia	Pectoralis and mammay aplasia: other muscles aplasia
Moderate mammary hypoplasia	Probable chest wall deformity	Always chest wall deformity
Light thoracic male asymmetry; female mammary asymmetry	Pronounced thoracic asymmetry	Major thoracic asymmetry in bot genders; mammary asymmetry due to aplasia in female

1.5. Diagnosis

The diagnosis of PS is not always apparent before adolescence, as it often becomes more evident as the patient becomes aware of asymmetry. The severe form usually does not go unnoticed in childhood. An adequate clinical diagnosis requires a physical examination of the thorax and both upper limbs, with measurements of each limb's musculoskeletal elements and comparison with the opposite side. Even in mild forms, differences in dimensions are apparent. Complementary studies, such as ultrasound, mammography, X-rays, CT scans, MRI, and angiography, can aid in making a proper diagnosis by revealing various PS findings [9] [10].

1.6. Classification

Multiple classifications have been developed to determine the appropriate treatment of the thorax, breast, and/or upper limb and hand, depending on the degree of involvement. The surgical treatment must first be divided and determined by the areas to be treated. The primary objectives of reconstruction are aesthetic, with the exception of upper limb malformations that impair function, such as syndactyly. A range of surgical techniques, including breast prostheses, thoracic prostheses, free and pedicled flaps, and autologous adipose tissue grafts, may be employed and can be combined as necessary [11].

Poland Syndrome presents with a wide variability of thoracic anomalies and different surgical options, which led Romanini *et al.* to propose the thoracic, breast, and nipple-areola complex (TBN) classification. Thoracic anomalies are classified as follows: thorax (T), from T1 (muscle defect only) to T4 (complex deformity with rib and sternal involvement); breast (B), in B1 (hypoplasia) or B2 (amastia); and nipple-areola complex (N), from N1 (dislocation < 2 cm) to N3 (athelia) (**Table 2**) [12].

Table 2. TBN classification of thoracic anomalies in poland syndome (translation of Romanini *et al.*).

		Anomaly
T	Thoracic	
	T1	Hypoplasia or aplasia of pectoralis muscles and soft tissue
	T2	T1 and sternal deformity, pectus excavatum and/or carinatum
	T3	T1 and rib aplasia
	T4	T1, T2 and T3 (muscle, sternum, and rib defect)
B	Breast	
	B1	Breast hypoplasia
	B2	Breast aplasia
N	Nipple-areola complex	
	N1	NAC hypoplasia with dislocation of <2 cm
	N2	NAC hypoplasia with dislocation of >2 cm
	N3	Absent NAC
NAC*	Nipple-areola complex	

2. Research Theme

Poland Syndrome is an entity with a frequency that remains unknown in the Latin American population. However, its incidence can be estimated based on reports from Anglo-Saxon literature. A study conducted by Márquez and Barrera *et al.* at the Mexican Instituto Nacional de Pediatría over a 10-year period (2002-2012)

described an incidence of approximately 1 in 15,000, with a predominance of female patients. It is suggested that the deformity in men may go unnoticed or be subtle enough that patients do not seek medical evaluation.

In this article, we aim to describe the population diagnosed within the Petróleos Mexicanos health system, as it belongs to the largest parastatal company in the country and may reflect the country's prevalence of the disease.

3. Material and Methods

Objective:

This study aimed to determine the prevalence of Poland Syndrome among patients at the Central South High Specialty Hospital of Petróleos Mexicanos (PEMEX) and to compare the hospital's treatment algorithms with those reported in current literature. Additionally, the study sought to classify cases according to the TBN classification and describe the reconstruction methods employed.

Study Design:

A retrospective review of electronic medical records from 2013 to 2023 was conducted. All patients diagnosed with Poland Syndrome during this 10-year period were included in the study.

Data Collection:

The study reviewed 10 years of electronic medical records to identify cases of Poland Syndrome. Each case was classified according to the TBN classification, and the reconstruction methods used were documented.

Results:

Eight cases of Poland Syndrome were identified during the study period, with a significant predominance of female patients (7 females, 1 male), consistent with existing literature. Interestingly, 75% of the cases (6 out of 8) exhibited left-side involvement, which contrasts with the literature that typically reports a predominance of right-side presentation. Each case was classified using the TBN classification, and the corresponding reconstruction method was documented and analyzed.

4. Ethical Considerations

This research was conducted in accordance with the "Regulations of the General Health Law" concerning health research, specifically Title Two, Chapter I, Article 17, Section I, which pertains to research without risk and does not require informed consent. The data were collected from electronic records.

In carrying out this study, the "Declaration of Helsinki of the World Medical Association", which sets forth ethical principles for medical research involving human subjects, was not violated. This declaration was adopted at the 52nd General Assembly in Edinburgh, Scotland, in the year 2000. Furthermore, based on the amendment made in Tokyo in 1975, this study was reviewed and approved by the local research and bioethics committee of the Central South High Specialty Hospital, Petróleos Mexicanos.

5. Results

The most frequent thoracic anomalies were T1 (100%) and N3 (50%), similar to findings in worldwide literature. The surgical approach to thoracic reconstruction was based not only on the patient's age and sex but also on the type of anomaly according to the TBN classification. A two-step approach with a tissue expander was required in 90% of the patients with N2 and N3 anomalies; only two cases were resolved with a single-step reconstruction using a breast implant.

6. Discussion

The current understanding of Poland Syndrome has evolved since its initial descriptions. Diagnostic criteria now include the aplasia or hypoplasia of the pectoralis major muscle, along with at least one other ipsilateral anomaly on the affected side. These may include aplasia or hypoplasia of the pectoralis minor muscle, rib arches, breast (amastia), nipple (athelia), pectoral or axillary alopecia, hypotrophy of subcutaneous tissue and/or sweat glands, and hand malformations [13] [14].

The degree of dysfunction in Poland Syndrome is usually mild to moderate, and thus, the primary reason for surgical management is aesthetic. Cardiorespiratory dysfunction tends to present early in severe cases, although these are infrequent.

Surgical methods include tissue expanders, latissimus dorsi flaps, fat grafting, free microvascular flaps, and custom silicone prostheses. Among the more recent techniques is the use of a flap dependent on the omentum, obtained laparoscopically [15].

Options for surgical correction of the chest wall depend on the patient's age and sex, the presence of the latissimus dorsi muscle, and the severity of the thoracic deformity. In male patients, chest wall reconstruction can be performed from the beginning of adolescence; in female patients, intervention can be considered after the completion of breast development [16]. An expander can be placed in the subcutaneous tissue on the affected side, which must later be removed for breast reconstruction using an ipsilateral latissimus dorsi myocutaneous flap. If the latissimus dorsi muscle is absent, the rectus abdominis muscle or the contralateral latissimus dorsi muscle can be used instead [17].

These techniques have achieved excellent results in breast volume reconstruction; however, one of the main challenges of the surgery is the reconstruction of the anterior axillary pillar and the filling of the infraclavicular fossa (**Figure 2**).

7. Conclusions

Poland Syndrome is a rare and heterogeneous congenital malformation characterized by hypoplasia or aplasia of the pectoralis major muscle and other associated anomalies. Its etiology remains unclear, although vascular theories dominate current hypotheses. Despite the variation in presentation and severity, surgical intervention is often warranted, primarily for cosmetic reasons [18].

The cases identified in this study, although small in number, demonstrate the diversity of PS presentations. The outcomes highlight the importance of individualized

surgical planning, considering factors such as the extent of deformity, patient sex, and age.

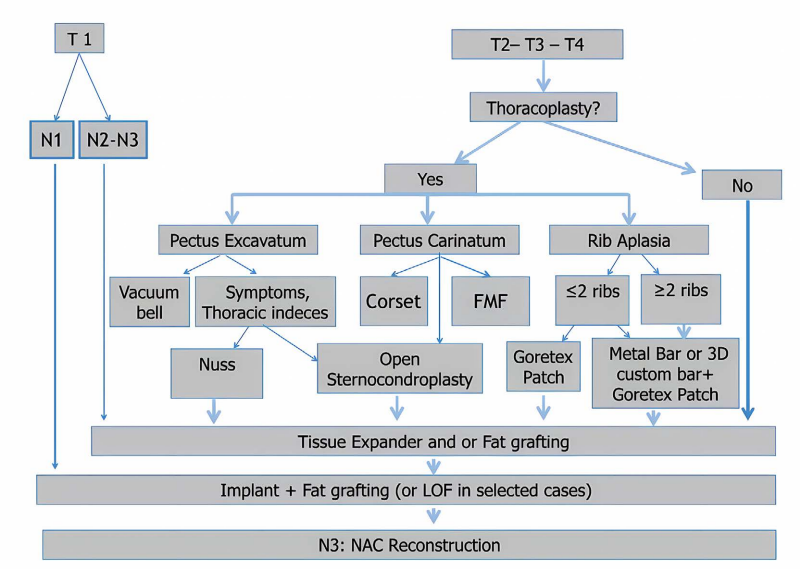

Figure 2. Algorithm for treatment of Poland syndrome.

Given the rarity of this syndrome, it is essential to continue accumulating case studies and research to refine diagnostic criteria and improve surgical techniques, ensuring better outcomes for patients with Poland Syndrome (**Figure 3** & **Figure 4**).

Figure 3. 19 years old female with PS. CT scan with 3D reconstruction of bone, muscle and soft tissue. Mammary agenesis and atelia, ipsilateral upper limb hypoplasia is observed as well as sternocostal hypoplasia with thorax deformity and pectoralis minor and major muscle aplasia.

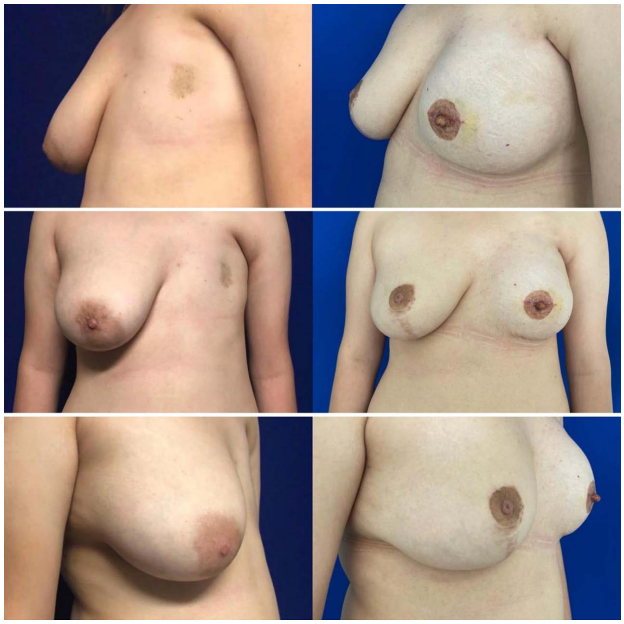

Figure 4. 19 years old female with PS preoperative and postoperative results with 3 steps reconstruction with breast expander and posterior change for breast implant with NAC reconstruction and areola pigmentation.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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