CASE REPORT

Management of Chest wall Deformity in Poland's Syndrome- A Case Report

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ABSTRACT

Background: Poland's syndrome is a rare congenital condition characterised by unilateral chest wall hypoplasia or aplasia, often accompanied by ipsilateral upper limb abnormalities. Limb anomalies may include brachysyndactyly or underdeveloped arm structures. Advancements in surgical techniques and a multidisciplinary approach have significantly improved outcomes for patients with Poland syndrome.

Case details: A 21-year-old male presented to the Plastic Surgery outpatient department with a complaint of left-chest wall deformity. On physical examination, the left anterior chest wall was depressed, and the nipple was displaced. The absence of the pectoralis major sternocostal head was visible during shoulder abduction. No signs of ipsilateral digital abnormality were noted. The computed tomography scan reported a complete non-visualisation of the left-sided pectoralis major and minor muscles. Hence, a diagnosis of Poland's syndrome involving the left hemithorax in a male patient was established. The patient opted to have reconstructive surgery for purely cosmetic reasons. Ipsilateral latissimus dorsi muscle transfer was done.

Conclusion: Autologous reconstruction with ipsilateral latissimus dorsi muscle transfer for chest deformity provides durable and aesthetic results for patients with Poland's syndrome, addressing both the functional and cosmetic aspects of the condition.

Keywords: Poland syndrome, latissimus dorsi, chest wall deformity

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INTRODUCTION

Poland Syndrome is a rare congenital condition characterised by the underdevelopment or absence of the pectoralis muscle on one side of the chest, leading to chest asymmetry. It may also involve other abnormalities like underdeveloped breast tissue, rib deformities, and hand anomalies like brachysyndactyly. The severity of the condition varies, and treatment often involves surgical correction and physical therapy to address functional and cosmetic concerns.

The surgical correction of Poland's syndrome chest wall deformities depends on age, sex, and severity. Severe cases with rib aplasia may require staged procedures with initial rib reconstruction during childhood, followed by muscle flap transposition and breast augmentation after puberty in females. For mild deformities, surgery is typically deferred until post-puberty. Techniques include rib grafts, prosthetic mesh, and muscle flaps, such as latissimus dorsi transfer, to stabilise the chest and restore symmetry. Advanced imaging aids in planning, especially for hypoplastic muscles. This is a case report on a variant of Poland's syndrome involving the left side chest wall of an adult male patient. He underwent the single-stage autologous reconstruction of chest wall deformity.

CASE DESCRIPTION

A 21-year-old male presented to the Plastic Surgery outpatient department with a complaint of left-sided chest deformity since birth. On physical examination, the left

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anterior chest wall was depressed. Physical examination revealed the absence of the pectoralis major sternocostal head during shoulder abduction (Figure 2) with no associated hand abnormalities. The computed tomography scan (CT scan) reported the absence of pectoralis major and minor muscles on the left side, and the anterior aspects of the left third and fourth ribs were hypoplastic, not articulating with the sternum (Figure 1). Hence, a diagnosis of Poland's syndrome involving the left hemithorax in a male patient was established.

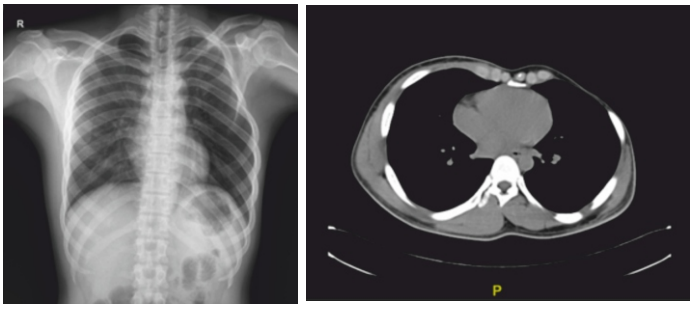


Fig. 1: A- Absent anterior part of left 3,4 ribs. B-Absence of pectoralis major and minor muscles on the left side. Anterior aspects of the left third and fourth ribs are hypoplastic, not articulating with the sternum



Fig. 2: A and B show the preoperative chest wall deformity; C and D show the 6 monthly postoperative pictures. The anterior axillary fold is well formed and the contour over the lateral chest wall improved on the left side

The ipsilateral latissimus dorsi (LD) muscle transfer was planned. The patient was positioned in the right lateral position. Under aseptic precautions and general anaesthesia, parts were painted and draped. The anterior border of the LD muscle was marked. Tumescent infiltration was given along the borders. A longitudinal exploratory incision was made, and the muscle was identified. The ipsilateral LD and serratus anterior muscles were also hypoplastic. LD muscle was freed around the anterior, inferior, and posterior borders with 1cm of fascia and transferred with the tendon as the pivot point. The patient's position was subsequently changed to supine. A subcutaneous anterior pocket was created into which the muscle was transposed (Figure 3). The distal end of the muscle was fixed to the skin using bolster sutures. The donor site was closed after the suction drain was placed.

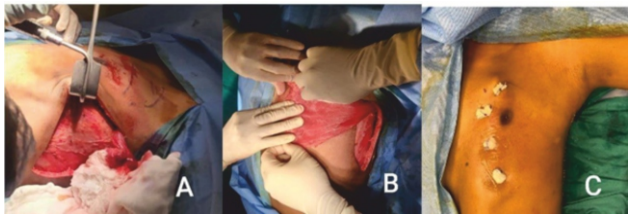


Fig. 3: A- A subcutaneous pocket in the anterior chest wall is created. B- Latissimus dorsi muscle flap is raised and transposed anteriorly. C - Muscle flap is anchored to the skin of the anterior chest wall with bolster sutures.

The patient was followed up regularly, and the chest wall depression on the left was remarkably reduced on the lateral aspect, and the anterior axillary fold was well defined. Even though the anterior chest wall deformity was not fully corrected, the patient was satisfied with the outcome. The patient did not complain about any reduction in shoulder range of movement.

DISCUSSION

Poland's syndrome was first described by Alfred Poland in 1840. The aetiology remains unknown. It is rarely genetically inherited and is regarded as a sporadic event.[1] Poland's syndrome is characterized by hypoplasia of the breast and nipple, scarcity of subcutaneous tissue, absence of the costosternal portion of the pectoralis major muscle, lack of the pectoralis minor muscle, aplasia or deformity of the costal cartilages or ribs II to IV or III to V, alopecia of the axillary and mammary region, and unilateral brachysyndactyly.[2] Clinical manifestations of Poland's syndrome are extremely variable.[3] The reported incidence of Poland's syndrome ranges from 1 in 7,000 to 1 in 100,000. Males are affected more frequently by a 2:1 to 3:1 ratio.[4-7] Poland's syndrome has also been diagnosed in 1 of 19,000 mammograms.[7] The right side of the body was found to be involved in 60% to 75% of patients. It has been suggested that during the sixth week of gestation, the injury

occurs due to a regional vascular defect of the subclavian artery. It is a period associated with the splitting of the two heads of the pectoralis major and the development of tissues between the digits. Another hypothesis is that disruption of the lateral plate mesoderm (from which the pectoralis muscle develops) between 16 and 28 days after fertilisation may account for all the defects.

In some cases of Poland's syndrome, the latissimus dorsi, external oblique, and serratus anterior muscles are also affected. Whenever a well-defined axillary fold, composed of the lower border of the pectoralis major muscle, is absent, the posterior fold, formed by the latissimus dorsi and teres major muscles, can be seen from the front. The unilateral chest wall deformity is often due to hypoplasia or aplasia of ribs and cartilages, particularly ribs II-IV or III–V. The sternum may rotate toward the affected side, and severe chest wall depression occurs in 11–25% of cases.[8-10] Lung herniation occurs in 8% of cases, with paradoxical chest wall motion and reduced vital capacity. Rare complications include contralateral lung bulla and heart displacement. Unilateral hand anomalies vary from syndactyly to ectrodactyly, occurring in 13.5–56% of cases. Variants of this syndrome include isolated pectoral hypoplasia or "partial Poland's sequence," which is more common than full manifestations. Poland's syndrome coexists with Möbius syndrome (15% of cases), Klippel-Feil syndrome, renal anomalies (e.g., agenesis or duplication), Sprengel's deformity, and certain cancers (e.g., leukaemia, breast cancer).[11-14]

Operative indications include asymmetry and depression of one side of the chest wall, with the potential for worsening over time, insufficient protection for internal organs, abnormal chest wall motion during breathing (paradoxical movement), underdeveloped or absence of the breast in females, and aesthetic concerns.

The surgical approach is tailored to the degree of deformity, age, and sex. Repairs can be staged or performed in a single operation, depending on severity and patient factors. In adults, single-stage stabilisation and reconstruction are common. The procedures may include rib grafts, muscle transposition, and, for women, augmentation mammoplasty. In severe rib cage defects, rib grafts, mesh, or a combination are done. Grafts are secured to the sternum and ribs, often corrected with osteotomy. The milder forms are managed by splitting normal ribs to bridge gaps or reattaching fused ribs. Muscle and breast reconstruction is by use of musculocutaneous flaps, such as latissimus dorsi or TRAM flaps, for better coverage and chest wall projection. A musculocutaneous flap is preferred over a simple muscle flap due to the thinness of the skin and subcutaneous tissues, which are often insufficient to support a substantial volume of transferred muscle and prosthetic material. Additionally, transferring a latissimus dorsi musculocutaneous flap addresses both the loss of the anterior axillary fold and the associated muscle mass. However, this approach has drawbacks, including the creation of a secondary scar and the removal of a major shoulder and arm muscle. The condition of the latissimus dorsi muscle significantly impacts surgical planning and outcomes. In some cases, if the latissimus dorsi is underdeveloped or absent, microsurgical transfer of the contralateral latissimus dorsi is a viable alternative. Other options include upper gluteal flaps or microvascular-free transverse rectus abdominis muscle (TRAM) flaps. Preoperative angiography is crucial for evaluating vascular supply and ensuring the success of microvascular tissue transfers.[15] In females, augmentation of hypoplastic breasts after puberty is advisable for symmetry. Advanced techniques include microsurgical transfers or prefabricated implants and endoscopic-assisted techniques for minimising scars over the chest wall. Silicone implants are commonly used in Poland's syndrome to correct chest wall and breast asymmetry, particularly in post-pubertal females. They are placed under the skin or muscle flaps to enhance contour and projection, often combined with other reconstructive techniques like rib grafts or muscle transposition for better outcomes. Due to thin overlying tissues, implants may require musculocutaneous flaps for support, and long-term monitoring is essential to address potential complications.

CONCLUSION

Poland's syndrome is an uncommon congenital musculoskeletal anomaly with a significant spectrum of presentations. It is classically characterised by aplasia or hypoplasia of the chest wall muscle and is variably associated with ipsilateral limb anomalies. Mostly, Poland's syndrome is treated surgically with a strong emphasis on aesthetics. The chest wall may remain flat without muscle flaps. Postoperative physiotherapy can enhance results by hypertrophying transferred muscle flaps. Careful

preoperative imaging and planning improve outcomes. Early correction ensures stability, while staged procedures align with growth and development.

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