Clinical Analysis of 113 Patients With Poland Syndrome

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Background. **Poland syndrome is a rare congenital anomaly characterized by the partial or complete absence of pectoral muscles, varying thoracic deformities, and hand anomalies. To date, many variants of this syndrome and its accompanying anomalies have been reported.**

Methods. **In our clinic, 113 patients were diagnosed with Poland syndrome between 1990 and 2014. A latissimus dorsi muscle transfer was performed on 6 of these patients.**

Results. **Out of 113 patients, 63 (55.7%) were diagnosed with the syndrome on the right side, 42 (37.1%) were diagnosed on the left side, and 8 (7%) had a bilateral diagnosis. The partial or complete absence of the pectoralis major muscle was detected in all patients. Although 81 (71.6%) patients had a complete absence of the pectoralis major muscle, 32 (28.3%) were lacking only the sternocostal head of the muscle. In the analyzed cases, Poland syndrome was also found to be accompanied by specific anomalies. The most common anomaly accompanying Poland syndrome in these patients was Sprengel deformity, seen in 18 patients. Symmetry and stabilization of the chest wall were performed in 6 patients through transfer of the latissimus dorsi muscle.**

Conclusions. **Poland syndrome is a rare congenital anomaly, which has several variants and accompanying anomalies. The absence of several muscles in addition to the pectoral muscle can be seen in patients with Poland syndrome. Sprengel deformity is the most common accompanying anomaly. Several surgical procedures have been reported for the syndrome; for example, transposing the latissimus dorsi muscle is an effective procedure in terms of stabilizing the chest wall and providing optimum symmetric body appearance.**

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Poland syndrome is a congenital syndrome characterized by agenesis of the pectoralis major muscle, absence or hypoplasia of the pectoralis minor muscle, a varying number of rib defects, anomalies of the breast and nipple, a lack of subcutaneous tissue, the absence of pectoral and axillary hair, and hand anomalies (Fig 1A, 1B, 1C). The first case was reported by Alfred Poland in 1841. Based on this report, the first definition of the syndrome was made in the English-language literature [1]. Since then, many components and accompanying anomalies of the syndrome have been reported (Fig 1). The absence of the pectoralis major muscle is the most common component of Poland syndrome [2]. To be diagnosed with Poland syndrome, patients must present with 1 or more of the other components in addition to the affected pectoral muscle [2–5]. It is rare to have a case that includes all clinical indicators. The incidence of the syndrome is reported to be between 1:10,000 and 1:100,000. The syndrome usually manifests as sporadic, and the percentage of patients with a familial connection is less than 1%. Although it usually presents as unilateral, a low proportion of patients manifest bilateral symptoms [2, 3]. Common complaints from patients are related to asymmetric body posture. Therefore, the aim of surgical treatment is a symmetrical and stable chest wall.

Because Poland syndrome is rare, reports on the syndrome generally present individual cases. Published case series are rare. Our study of 113 diagnosed patients represents the largest unified analysis of the syndrome; thus, this case series is a significant contribution to the literature.

Patients and Methods

This study was approved by the GMMA Haydarpaşa Training Hospital Institutional Review Board. We retrospectively searched the institutional thoracic surgery database to identify all patients who had a diagnosis of Poland syndrome between January 1990 and June 2014. Patients with isolated pectoral muscle agenesis were excluded from the study; only patients with at least 2 manifestations of the syndrome in addition to pectoral muscle agenesis were included. We identified 113 patients with a diagnosis of Poland syndrome. The first 28 patients had been previously reported [6]. All patients were male except for 1, and the average age was 21.2 years (range, 6 to 38 years). Only 5 (4.4%) had a family history of the syndrome. A detailed physical examination was performed on all patients, and all patients underwent complete blood counts, routine biochemical tests, and posteroanterior and lateral chest x-rays. Patients with

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Fig 1. (A) The appearance of a patient with left-sided Poland syndrome (agenesis of the pectoral muscles, hypoplasia of the breast and nipple, hypoplasia of the subcutaneous tissue, and upper extremity shortness). (B) The appearance of a patient with right-sided Poland syndrome and Sprengel deformity. (C) The appearance of hands of a patient with left-sided Poland syndrome and brachysyndactyly.

hand anomalies were evaluated with hand x-rays. Possible accompanying anomalies were investigated by a thoracic computed tomographic (CT) scan and an abdominal ultrasound examination. Patients with cardiac complaints and dextrocardia were evaluated by an echocardiograph.

Surgical treatment for symmetry and stabilization of the chest wall was recommended for 14 patients who had large rib cage defects or lung herniation. Only 6 patients accepted the offer. During surgical treatment, the latissimus dorsi muscle flap was transferred to the pectoral region to restore the contour deformity due to the absence of the pectoral muscle and ribs.

The procedure for the surgery is as follows. A template of the pectoral region deformity, emphasizing its volume deficiency, was drawn on an acetate film sheet. The acetate template was used to mark the designated harvesting borders of the latissimus dorsi muscle on the skin of the thorax. While determining the muscle harvesting borders, an additional 5 cm of muscle segment was included around the muscle to take into account the contraction of the muscle after detaching. Before starting the surgery, a 10×8 cm fusiform area, corresponding to the most significant volume-deficient area of the template, was also marked on the skin. Together, the subcutaneous fat tissue and the underlying muscle are intended to yield extra volume in this fusiform area.

In the lateral decubitus position, the anterior border of the latissimus dorsi muscle was exposed after a 15-cm vertical skin incision on the posterior axillary line. An additional 10-cm horizontal skin incision toward the spine was also made from the caudal end of the vertical incision. The muscle dissection was held through a loose avascular plane under the subcutaneous tissue and the skin was marked on the area under which the subcutaneous fat would be included. The dissection was done at a more superficial layer, namely under the dermis, at the marked skin area to incorporate the subcutaneous fat with the muscle for a larger volume. The skin was not included to the harvested flap. The thoracodorsal artery pedicle of the muscle was dissected up to the subscapular artery, and the thoracodorsal artery, comitantes veins, and thoracodorsal nerve were preserved intact during the dissection. The latissimus dorsi muscle insertion on the humerus was also preserved. The marked harvest borders at the anterior, posterior, and caudal parts of the muscle on the thorax were incised to release the muscle attachments from the spinous processes of vertebra, thoracolumbar fascia, iliac crest, ribs, and scapula.

The latissimus dorsi muscle flap was harvested with its subcutaneous fat tissue on its thoracodorsal neurovascular pedicle (Fig 2A). After the flap was completely released from the thorax, a subcutaneous tunnel was created from the donor site toward the pectoral region and sternum. The latissimus dorsi muscle flap with its overlying subcutaneous fat tissue was transferred to the pectoral region through the dissected subcutaneous tunnel (Fig 2B). The anterior border of the muscle was sutured to the clavicle through a skin incision made under the clavicle. The caudal and posterior borders of the latissimus dorsi muscle flap on the thorax were sutured to the sternum and ribs with stitches in a way that imitated the orientation of the pectoralis major muscle. The subcutaneous fat tissue over the latissimus dorsi muscle provided the extra volume under the pectoral region skin to restore the contour defect. The postoperative view of