

POLAND SYNDROME: TREATMENT AND MANAGEMENT



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Poland Syndrome is a very rare congenital anomaly that can be characterized as the partial or complete absence of pectoral muscles. It is generally presented with many coexisting thoracic anomalies. (1-4)

Although the title -Poland syndrome- has been used for referring to different combinations of pectoral muscle deficiencies, syndactyly and chest deformities, various variants and accompanying diseases have been reported over time. Even though there are few case series reported in the literature, the largest single-center series had been published in our country. (1) Due to its rarity publications are generally in the form of case reports. The major component seen in patients with Poland syndrome is complete or partial absence of the major pectoral muscle. In addition to that patients having one or more of the other components are diagnosed with Poland syndrome. (5,6)

The treatment of Poland syndrome aims to provide an aesthetic and symmetrical appearance to the patient. There are multiple different options as bone grafts, patches, prosthetic materials, muscle flaps and combinations of these methods are used for the correction of the deformity.

HISTORY

During a routine dissection of a 27-year-old cadaver of a prisoner in 1841, Sir Alfred Poland had discovered the fact that left pectoralis major

and minor muscles together with the serratus anterior muscle were partially absent. Although two case reports including one case in each was reported in 1826 and 1839, these publications unfortunately could not be able to draw any attention at the time (3,7,8).

Following the first definition of Poland syndrome, many other different components had been reported as part of the syndrome. Thomson made the full description of the characteristics of the syndrome in 1895 (1-3). Dr. Furst published the first report regarding etiology and declared the hypothesis with the involvement of embryonic factors (9). Patrick Clarkson reported Poland syndactyly in 1962, referring to a case similar to the patient reported by Alfred Poland. Although many names had been attributed to the disease over the years, Baudinne et al. used the term Poland syndrome for the first time. (2,3)

The first report on the surgical treatment of Poland syndrome was published by Ravitch and Hansdelsman in 1952 (2). Tansini is the surgeon who performed the first myocutaneous flap surgery in the history in 1896 which is one of the most frequent options for the treatment of Poland syndrome (10). On the other hand the silicone pectoral implant, which has been preferred more frequently today, was first reported by Horn and Aiache in 1990 (11).

The number of case series published about Poland syndrome is very few. Dr. Bing reported his

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ternative is to place the prosthesis in an acellular dermal matrix. (36) In cases where the skin cannot be adequately expanded before the silicone prosthesis, muscle transfer should be performed in the form of a musculocutaneous flap. Muscle transfer should be done later in a second surgery in order to avoid atrophy in those patients in who tissue expanders had been used. (36)

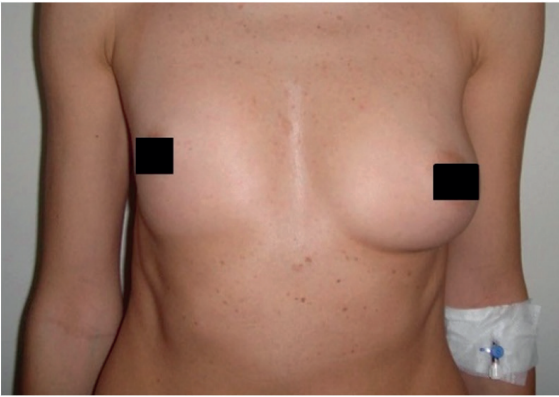


Figure 11 (a): Breast asymmetry in a patient with right-sided Poland Syndrome



Figure 11 (b): Application of bilateral silicone prosthesis in order to maintain symmetry.

In some cases, the opposite side can be supported with a small prosthesis to ensure optimum symmetry (Fig. 11 b). If breast sagging exists on the non-affected side of the chest, mastopexy (breast lift) has to be performed. On the other hand, if the breast is hypertrophic, it is appropriate to choose reduction mammoplasty as a surgical option. Patients may have areola and nipple involvement in a synchronous manner. In these patients, symmetry can be achieved by removing

the skin circularly including a part of the areola on the healthy side in order to provide a symmetrical appearance, by tattooing if there is no areola on the affected side, and by performing nipple reconstruction or reduction. (37)

The procedure to be performed to provide breast and nipple symmetry should be determined specifically for the anomalies the patient has. Surprisingly, there has been a publication reporting gynecomastia on the unaffected side in a patient with Poland syndrome. The body symmetry of this patient was achieved by performing gynecomastia correction surgery. (38)

In conclusion, Poland syndrome can be described as the absence of unilateral pectoral muscle accompanied by one or more ipsilateral anomalies related with the thoracic cage or extremities. The components participating in the syndrome determine the condition of the diagnosed patient and the plan to be followed in the treatment. Aesthetic concerns build up the mainstay of the patients' complaints. The primary goal in the treatment strategy should be to protect vital organs, especially the lungs and heart. The main purpose of the treatment is to provide the stability to the rib cage and the maximum symmetry to the body appearance.

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