Correction of Poland's Syndrome: Case Report and Review of the Current Literature

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Keywords

Poland's syndrome · Breast hypoplasia · Breast reconstruction · Malformation

Summary

Background: Poland's syndrome, a rare congenital anomaly, consists of unilateral absence of the pectoralis major muscle, ipsilateral brachysyndactyly, and occasionally associated other malformations of the anterior chest wall, mamilla, and mamma. **Case Report:** In the case of a 32-year-old woman, marked hypoplasia of the right breast and the right nipple, malformation of the right upper limb with brachysyndactyly and microdactyly were noted, all present since birth. This paper describes the surgical technique and possible complications of reconstruction of the chest wall deformity with a solid silicone implant and a latissimus dorsi flap. The current literature on the topic is reviewed. **Conclusion:** The surgical method with autologous material and implants is a sufficient technique for chest wall reconstruction and gives a good long-term result in case of Poland's syndrome.

Schlüsselwörter

Poland-Syndrom · Brusthypoplasie · Brustrekonstruktion · Malformation

Zusammenfassung

Hintergrund: Das Poland-Syndrom, eine seltene kongenitale Anomalie, ist durch eine einseitige Fehlbildung des Mamillen-Areola-Komplexes und der Mamma sowie des Musculus pectoralis major, eine ipsilaterale Brachysyndaktylie und gelegentliche Fehlbildung der Brustwand gekennzeichnet. **Fallbericht:** Bei einer 32-jährigen Frau wurde eine ausgeprägte Hypoplasie der rechten Mamma und des Mamillen-Areola-Komplexes sowie eine Fehlbildung des rechten Armes mit Brachysyndaktylie und Mikrodaktylie festgestellt, die alle seit ihrer Geburt vorlagen. Dieser Artikel beschreibt die operative Therapie und die möglichen Komplikationen, die bei der Korrektur mittels einer Latissimus-dorsi-Lappenplastik und einer Implantateinlage auftreten können. Eine Zusammenfassung der aktuellen Literatur wird geboten. **Schlussfolgerung:** Die Methode der autologen Mamma-rekonstruktion mit Implantateinlage ist eine zuverlässige Methode zur Brustrekonstruktion bei Patientinnen mit Poland-Syndrom und ist mit einem guten Langzeitergebnis verbunden.

Introduction

Poland's syndrome was initially reported by Sir Alfred Poland in 1841 as an absence of the pectoralis major and minor muscles and malformation of the ipsilateral upper limb [1]. Poland's syndrome is also described as Poland's syndactyly or 'Amazone syndrome'. Other authors characterized the syndrome by congenital aplasia or hypoplasia of the sternocostal head of the pectoralis major muscle, associated with ipsilateral malformation of the breast, nipple, and the upper limb. The incidence has been estimated between 1:30,000 and 1:80,000 of live births, with a gender predominance in men at a

ratio of 3:1 [2]. At present, approximately 400 cases of Poland's syndrome have been reported. The literature describes abnormalities associated with the syndrome, including aplasia or hypoplasia of the rib cage, vertebral defects, and dextrocardia as common findings. In 4 cases of the literature, kidney abnormalities associated with the syndrome have been reported [3].

The cause of Poland's syndrome is unknown. However, interruption of the early embryonic blood supply in the subclavian arteries, the vertebral arteries, and/or other branches has been hypothesized. Most reported cases are sporadic, but the disease may be inherited as an autosomal-dominant trait. The responsible gene has not yet been mapped [4].

Fig. 1. Poland's syndrome preoperatively.

Fig. 2. Brachysyndactyly in Poland's syndrome.

Fig. 3. Intraoperative muscle flap to correct the chest wall defect in Poland's syndrome.

Fig. 4. Poland's syndrome postoperatively.

Several articles have reported on different kinds of neoplasms in Poland's syndrome patients and the association with an increased incidence of malignancy, such as leiomyosarcoma, neuroblastoma, leukemia, and Wilms' tumor. Summarizing 400 described cases of the syndrome, an increased risk of malignancy of the breast in this group of patients may be possible [5]. 5 cases of breast cancer have

been reported previously [6, 7]. The hypoplastic breast should be monitored similarly to the normal breast. There is no difference in diagnostics or therapy for breast cancer in these patients.

Case Report

A 32-year-old woman presented at the University of Halle, Germany, as an outpatient asking for reconstruction of her hypoplastic breast. She was born with Poland's syndrome, presenting with hypoplasia of the right breast and the nipple-areola complex and absence of the pectoralis major muscle (fig. 1). The ipsilateral upper limb showed brachysyndactyly (fig. 2). Ultrasound of the abdomen showed a normally configured urogenital tract. Size and position of both kidneys were correct. There was no sign of dextrocardia, hypoplasia of the forearm, thoracic cage defects, or herniation of the lung. She was within normal limits in height, weight, and intelligence.

Reconstruction was performed under general anesthesia with a skin-free latissimus dorsi flap (LD flap) and placement of a 215-ccm silicone implant. The incision was marked preoperatively for optimal positioning of the flap. The initial procedure was combined with contralateral mastopexy for symmetry (fig. 3). The flap was initially denervated to exclude contractions of the reconstructed breast. A small suction drain was placed alongside the implant.

The patient had an uneventful postoperative course (fig. 4). Postoperatively she obtained a symmetric result. The patient was lost to long-term follow-up.

Table 1. Publications on correction of Poland's syndrome, reported with number of patients operated, procedures done, and outcome

Author	Number of patients who received surgery	Surgery applied	Complications described	Of note
Fijałkowska and Antoszewski [8]	n = 66 retrospective cases, from 1 institution (58 female, 8 male pts)	47 with implant reconstruction, 9 with additional correction of healthy breast; 2 with expander	not described	pts received surgical correction when > 18 years of age; from Poland
Gascoigne and Malata [9]	n = 3 cases	20 years after previous implant reconstruction: now rupture suspected and contractures Baker III–IV: capsulectomy	1 with capsule adherent to chest wall – developed intraoperative pneumothorax, complete recovery; 2 with implant exchanged, pt developed fistula from implant cavity to pleural cavity, pt wished implant permanently removed; 3 with uneventful implant exchange	specific problems of implant exchanges for corrected Poland's syndrome; from England
Papadopoulos et al. [10]	n = 32 cases	16 women who had pedicled LD flaps, 12 with tissue expander or silicone implants, 4 who had free TRAM flaps	satisfaction maximum for LDM	QoL questionnaire, good long-term results; from Germany
Baratte et al. [11]	n = 9 cases	grade I, breast implant; grade II LD flap combined with a breast implant; grade III, breast and chest reconstruction	not presented	from France
dos Santos Costa et al. [12]	n = 15 cases	all had transposition of the omentum (omentum flap)	no severe postoperative complications	from Brazil
Fekih et al. [13]	n = 8 cases	3 implants, 1 autologous fat, 3 contralateral lipoaspiration, 1 contralateral breast resection	no intraoperative or postoperative complications occurred	from Tunisia
Borschel et al. [14]	n = 29 cases	male pts: 8 with custom chest wall implant; female pts: 4 with LD flap + implant, 16 with expander/implant, 1 with chest wall implant + breast implant	male pts: 2 with seroma; female pts: 1 with implant migration	from USA
Freitas Rda et al. [15]	n = 18 cases	5 LD flap + implant, 2 expander technique, 4 implants, 7 contralateral adjustment	not reported	from Brazil
Foucras et al. [16]	n = 27 cases	male pts: 2 with LD flap, 6 with implant; female pts: 9 with breast implant, 4 with expander, 4 with breast and thoracic implant, 2 with LD flap	65% good results, 24% medium results, 12% bad results; results worse in higher-stage cases	from France

Pt = Patient, TRAM = transfer of the rectus abdominis muscle, LDM = latissimus dorsi muscle, QoL = quality of life, LD flap = latissimus dorsi flap.

Review of the Literature

The PubMed database was screened for relevant articles within the last 10 years on 'Poland syndrome breast', reviewing publications that describe surgical procedures. Articles mentioning ≥ 3 cases are presented in table 1.

The type of surgery used depends highly on the extent of the malformation and individual patient preferences (e.g. choice about sole implants vs. LD flap). Newly described methods include omentum flap techniques.

Complications described in the literature are few and do not differ from complications seen in the respective procedures done for other reasons. Due to aplasia of muscular and fatty structures as well as rigid skin, contractures after implant insertion may be more frequent and should be treated with caution (beware of nearby pleura). Due to the congenital nature of the abnormality, patients may be treated early in life. Therefore, a future change in the figure of the patient has to be anticipated.

Conclusion

Our patient shows characteristic findings of Poland's syndrome including the absence of the right pectoralis major muscle and ipsilateral hand deformities.

Reconstruction of the chest wall in patients with Poland's syndrome may lead to intra- or postoperative complications [17]. If the skin cover of an implant is not flexible enough, it will not allow stretching in a single stage and it will be neces-

sary to first implement an expander. Other complications could be due to the absence or maldevelopment of the shoulder girdle muscles, e.g. the pectoralis, latissimus dorsi, trapezius, or serratus anterior muscles [18, 19]. In case of an ipsilateral latissimus dorsi muscle defect, reconstruction of the chest wall and microsurgical or pedicled transfer of the rectus abdominis muscle (TRAM) might be the best choice for reconstruction. If the axillary vascular system is defective, the internal mammary vessels are used as recipients for free TRAM reconstruction.

In the literature, other techniques, e.g. laparoscopic reconstruction using the omentum flap technique or autologous fat injection, are described [12, 20].

In summary, reconstructive surgery depends on the severity of the malformation. A hypoplastic breast may be reconstructed with an implant (possibly expansion first). In case of maldeveloped muscles (e.g. the pectoralis muscle), an LD flap seems indicated. In case of additional thoracic malformations, more extensive, individual surgery is needed. Additionally, the position of the nipple-areola complex may need correction. For symmetry reasons, surgery to the contralateral breast can be considered. Reconstructive surgery in patients with Poland's syndrome is feasible and recommended for psychosocial reasons in these patients.

Disclosure Statement

All authors declare that they do not have any competing interests.

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