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Integrated algorithm for reconstruction of complex forms of Poland syndrome: 20-year outcomes

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KEYWORDS

Poland syndrome;
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Pectoralis major;
Breast reconstruction

Summary *Background:* The extent of breast and chest-wall deformity varies widely in Poland syndrome, and a variety of techniques are required to correct them. The aim of this study is to analyse our experience and long-term follow-up, and to propose an algorithmic approach to this complex deformity.

Patients and methods: Retrospective case-note review from a prospectively maintained database was carried out on 37 patients. All patients treated between 1993 and 2013 were reviewed, and long-term outcome was assessed by an objective photographic documentation. The patients were invited to a photographic documentation, and the correlation between the subjective judgement and objective results in the late course was analysed. Based on our experience, we propose an algorithmic approach to this complex skeletal and soft-tissue deformity.

Results: Medical photographs were obtained from 32 patients (22 females and 10 males). The age range was 11–44 years. Significant reduction in asymmetry was possible in all patients. The mean follow-up time was 8.5 years (range, 2.2–20). The complex developmental defect involved the skin, subcutaneous tissue, breast, muscle, ribs and other skeletal abnormalities. The following treatments were applied alone or in combination: lipomodelling, liposuction, tissue expansion and implants, custom-made prosthesis, islanded pedicled latissimus dorsi muscle flaps, pedicled and free muscle-sparing transverse rectus abdominis flaps, prosthetic chest-wall reconstruction and various combinations including multistage procedures. Surgery on the contralateral breast in female and male patients was necessary in 22 patients (20 female and two male). Assessment of the late result by four non-professionals (two females and two males) and one male plastic surgeon showed that the objective aesthetic results of the authors' reconstructions according to the proposed algorithm were sufficient.

Conclusions: Poland syndrome is a complex developmental deformity in which several tissues are involved. It is crucial to have an armamentarium of techniques to tackle a variety of

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defects to meet patients' expectations, and to improve their quality of life. The clinical outcome of the currently presented algorithm proved beneficial in the decision process for the treatment of these congenital deformities.

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Introduction

Poland syndrome was described by Sir Alfred Poland as the absence of the sternocostal part of the pectoralis major muscle, the absence of pectoralis minor and serratus anterior muscles.¹ The original description comprised the mildest form. Clarkson rediscovered the anomaly and detected an association with the ipsilateral hand anomalies (sybrachydactyly).^{2,3} The incidence has been estimated to be between 1:30,000 and 1:80,000 of live births with gender predominance in men, with a ratio of 3:1.⁴ At present, approximately 400 cases have been reported. Following Poland and Clarkson, a variety of additional anatomical features have been described ranging from hypoplasia and aplasia of the nipple, subcutaneous fat deficiency, ipsilateral breast parenchyma deficiency as well as non-parenchymal tissue. The most severe form described included hypoplasia or aplasia of ipsilateral trunk muscles with skeletal deformities such as hypoplasia of the ribcage or absent sections of multiple ribs and scoliosis combined with deformities of the ipsilateral upper extremity.^{5,6} Poland syndrome represents an entity characterized by multiple tissue hypoplasia resulting in complex anatomical defect. It is imperative to consider an armamentarium of surgical strategies to improve those defects to the level of the patient's satisfaction. A variety of surgical techniques have been described in the literature over the last decades to address various deformities within the spectrum of Poland syndrome.^{7–11} These procedures have been mostly described as a single strategy for the reconstruction for various anatomical deformities.^{7–11} The aim of our study was to review our experience with the specific complex Poland deformities and our long-term outcomes, and to propose our algorithmic approach taking into consideration the anatomical defect, available donor sites and patients' personal circumstances.

Patients and methods

A total of 37 patients were treated by the senior author over the 20-year (1993 and 2013) period. Ethical review board approval was not required in our institution for this retrospective analysis when the project was started. Medical photographs of 32 patients were available for a retrospective observation period, comprising 26 females, one male-to-female transgender patient and 10 males. All patients were invited, and they presented themselves for photographic follow-up documentation. We obtained medical photographs from 32 patients. Retrospective case-note review was carried out on 37 patients. All patients

were reviewed and long-term outcome was assessed by objective photographic documentation, and the correlation between the subjective judgment and objective results in the late course were analysed. Standardized photographs of 32 patients were taken with the patient standing and their hands on each iliac crest. Photographs of frontal, oblique (left and right), lateral (left and right) and back views (if posterior chest-wall abnormality present or latissimus dorsi (LD) flap was planned to be used) were taken. The medical photographs were assessed by four clinically uninvolved professionals from specialist nurses and medical students to an independent plastic surgeon. All assessments were conducted individually, independently and anonymously. The photographs were scored on a visual analogue scale from 0 to 10 for shape and symmetry of the reconstructed components of Poland syndrome defects. Custom-made silicone prostheses were designed by the senior author, and they were manufactured by Nagor. Anatomical range of defects, areas requested by patients to be addressed, surgical techniques and, based on our experience, an algorithm for the management of the different deformities are presented.

Results

We treated 37 patients with Poland syndrome (26 females, one male-to-female transgender patient and 10 males). The mean age was 24.7 years ranging from 11 to 44. All patients underwent reconstruction of unilateral breast and/or chest-wall asymmetry due to a complex form of Poland syndrome. The mean follow-up was 8.5 years (range, 2.2–20). The complexity of anatomical defects associated with Poland syndrome in this series is presented in [Table 1](#). Patients' characteristics and the surgical procedures carried out in Poland syndrome patients are shown in [Table 2](#).

In the female group, the resultant congenital defects requiring correction included breast asymmetry in 100% ($n = 26$) of patients and one male-to-female transgender patient, correction of the anterior axillary fold 46% ($n = 17$), nipple reconstruction 5% ($n = 2$) and residual anterior chest-wall contour defect 5% ($n = 2$).

In the male group, all patients ($n = 10$) required correction of the anterior axillary fold and improvement of breast symmetry, and 5% ($n = 2$) required correction of the anterior chest-wall defect.

None of the patients with complex chest-wall hypoplasia (female or male) required the involvement of a cardiothoracic surgeon for chest-wall reconstruction to improve cardiovascular or respiratory problems.

In the female group, 27% ($n = 13$) required breast augmentation. Five per cent ($n = 2$) were previously

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