

A Rare Poland Syndrome Deformity: Humero-Pectoral Band

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Introduction

Since Dr. Lallemand has defined isolated pectoral muscle absence in 1826 [1], many articles have been published about Poland Syndrome. Ipsilateral syndactyly with pectoralis major muscle agenesis has been defined by Dr. Froriep in 1839 [1]. Dr. Alfred Poland has defined same entity in 1841 while he was a medical student [1]. Agenesis of the pectoralis major muscle's sternal head is the major finding in diagnosis of Poland Syndrome. Additionally, scoliosis, lung, rib cage (two to four or three to five costal cartilages), breast and nipple-areolar hypoplasia, ipsilateral brachysynphalangism, serratus, pectoralis minor, latissimus dorsi and external oblique muscles hypoplasia, absence of axillary hairs and sweat glands and decreased subcutaneous tissue would be seen together in Poland Syndrome [1, 2].

Case Report

A 2 year old boy was presented to our hand surgery team with the absence of pectoralis major muscle and a contracture like band between his right hemi-thorax and right humerus (Fig. 1). There was no associating thoracic or hand anomaly. His chest x-rays and magnetic resonance imaging findings support physical examination. There was no family history and this band disables his abduction and extension of the arm.

Five flap z-plasty has been designed for reconstruction of the axilla (Fig. 2). Flaps were elevated and fibrotic tissue was visualized (Fig. 3). This tissue was lying between anterior thorax wall and upper arm fascia. It was a firm structure with almost no vasculature over it. Total excision of the tissue for a better flap inset was planned. Excision of this tissue was done with unipolar cautery over pectoral and upper arm fascia. Then flaps' insets were done. This *five flap z-plasty* procedure let us to reconstruct anterior axillary fold and to treat the disability of the arm's extension and abduction movement (Fig. 4). Latissimus dorsi transfer to reconstruct pectoral region's concavity has not been planned because of the age of the patient.

Pathologic report of the specimen was reported as connective tissue.

Discussion

Poland Syndrome is a sporadic, congenital and unilateral disease which may be seen less than 1% in the same family [2]. It is more common in males (2:1-3:1) and frequently seen in right hemi-thorax (60–75%) [2]. The reported incidence of Poland's syndrome ranges from 1 in 7,000 to 1 in 100,000 [2]. Currently the prevailing theory in the

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Fig. 1 Fibrotic band which lies between anterior thoracic wall and humerus can be seen

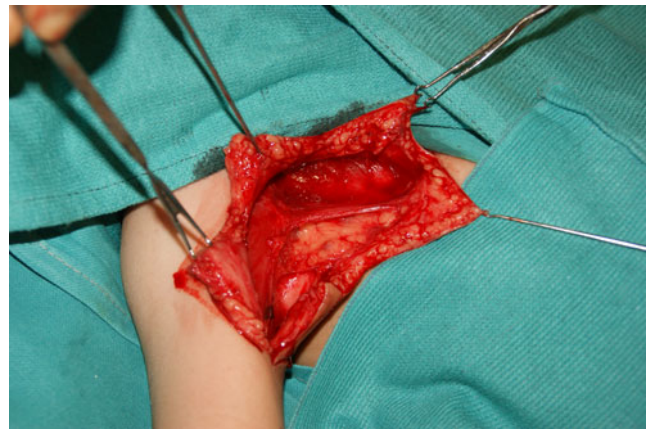


Fig. 3 The position of the band before it was being excised, just after the z-plasty flaps were elevated

etiology of Poland Syndrome is that, at the end of the sixth week of gestation, when the upper limb bud adjacent to the chest wall is still in a stage of development, the interruption of the embryonic blood supply causes hypoplasia of the ipsilateral subclavian artery or one of its branches [2]. Additionally, hypoplasia of the internal thoracic artery could cause the absence of the sterno-costal portion of the pectoralis major muscle, whereas hypoplasia of the brachial artery may lead to hand abnormalities [2]. Al-Qattan et al. have classified the hand anomalies of the Poland Syndrome in to seven types [3]. Presented case is accepted as type one according to this classification.

Four fundamental functions of z-plasty are to lengthen a scar, to break up a straight line, to move tissues from one area to another and to obliterate or create a web or cleft [4]. In this case we need all these functions in order to reconstruct anterior axillary fold. So a z-plasty design has been decided. However, for a better aesthetic result we need an advancement flap at the same time. We think that, if

there is supple skin which allows greater y-v advancement, it should be well used without any tissue lost. Tight bands would restrict this maneuver but like in this case, if excision of the band is possible, it should be done. Therefore, *five flap z-plasty*, in other words, *jumping man flap* has been preferred in this case because of its advantage of y-v advancement pattern over other z-plasty techniques. The main disadvantage of this technique is the midline split which narrows the vascular pedicle to the lateral flap tips. So their angles should be planned more obtuse (75°) than for a typical double opposing Z plasty.

Arsilan et al. have presented a case report in 2003 which has a similar humero-pectoral band. Additionally, their patient had a hand anomaly. They have treated this patient's thorax wall with a latissimus dorsi muscle flap. They have reported as this was the first description of this anomaly in the literature [5].

Recently, Seyfer et al. have published a report of 63 Poland Syndrome patient. They have presented a similar



Fig. 2 Design of the five flap z-plasty



Fig. 4 Post-operative 2 weeks result. Pre-operative limited abduction was well treated

case in their article with a humero-pectoral band, however they did not give any incidence for this entity [6]. They think that this would be a remnant pectoral muscle of which we think about the same. The number of published article about this “remnant” anterior axillary fold is very few [5, 6] although absence of anterior axillary fold is diagnostic for this syndrome.

Financial Disclosure Authors have no financial interest in any product mentioned in this study.

Ethical Standards This case report has been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki and its later amendments; also informed consent of the patient (and his family) has been taken.

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