DOI: 10.17816/PTORS5163-70

POLAND'S SYNDROME

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For citation: Pediatric Traumatology, Orthopaedics and Reconstructive Surgery, 2017;5(1):63-70

Received: 16.01.2017
Accepted: 22.02.2017

Poland's syndrome is a rare congenital condition classically characterized by partial or complete absence of chest muscles on one side of the body and usually webbing of the fingers of the hand on the same side. There may also be rib (aplasia or hypoplasia) and chest bone abnormalities, which may be noticeable due to less fat under the skin. Breast and nipple abnormalities may also occur, and underarm hair is sometimes sparse or abnormally placed. In most cases, the abnormalities in the chest area do not cause health problems or affect movement. Poland's syndrome most often affects the right side of the body and occurs more frequently in males than in females. The etiology is unknown; however, interruption of the embryonic blood supply to the arteries that lie under the collarbone (subclavian arteries) is the prevailing theory. There are many methods of operative correction because of the polymorphic clinical features of this syndrome. We gathered data on the etiology, pathogenesis, and clinical presentation of Poland's syndrome and reviewed the existing surgical treatment options.

Keywords: Poland's syndrome, aplasia (hypoplasia) of the pectoralis major muscle, chest wall deformity, brachydactyly, surgical treatment.

СИНДРОМ ПОЛАНДА

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Статья поступила в редакцию: 16.01.2017

Статья принята к печати: 22.02.2017

Синдром Поланда — редкий врожденный синдром, характеризующийся частичным или полным односторонним отсутствием большой грудной мышцы и врожденным пороком развития кисти со стороны поражения грудной клетки. Также могут быть аномалии ребер (аплазия или гипоплазия) и грудины (килевидная или воронкообразная деформация), которые отчетливо пальпируются ввиду истончения подкожно-жирового слоя. Часто при данном синдроме имеются аномалии сосково-ареолярного комплекса. Дефекты грудной клетки обычно носят косметический характер. Мужской пол превалирует над женским в соотношении 2–3 : 1, по частоте преобладает правостороннее поражение. На данный момент общепринятой теорией развития синдрома Поланда является нарушение кровотока по подключичной и/или позвоночной артериям и их ветвям на 6-й неделе внутриутробного развития. Ввиду полиморфности проявлений синдрома существует множество подходов к объему и срокам оперативной коррекции. В статье мы проанализировали данные по этиопатогенезу и клинической картине синдрома Поланда, а также по способам хирургического лечения.

Ключевые слова: синдром Поланда, аплазия (гипоплазия) большой грудной мышцы, деформация грудной клетки, брахидактилия, хирургическое лечение.

Introduction

Poland syndrome is a congenital syndrome characterized by unilateral (rarely bilateral) hypoplasia (aplasia) of the greater pectoral muscle (and possibly of the smaller pectoral muscle) and ribs (usually from 2 to 5), atelia (defect of the nipple-areolar complex), amastia (absence of breast tissue), thinning of the subcutaneous fatty layer of the chest, absence of hair in the axillary region and on the chest on the side of the lesion, as well as congenital malformations of the upper extremity, often in the form of symbrachydactyly.

There are two forms of the disease: complete and incomplete. A combination of chest and hand defects is complete Poland syndrome. Isolated hypoplasia of the greater pectoral muscle is diagnosed as an incomplete syndrome, and it occurs more often than the complete form [1]. According to Yiyit et al. (2015), the combination of one or more symptoms in conjunction with a lesion of the greater pectoral muscle is necessary for the diagnosis of Poland syndrome.

The incidence rate of the Poland syndrome is one case per 10–100 thousand people [2]. Poland syndrome more often affects males at a ratio of 2–3:1 [3]. The individual symptoms were first noted by Lallemand (1826) and Frorier (1839), with a complete description of Poland syndrome provided by the English medical student Alfred Poland in 1841 [4]. In 1895, Thompson summarized all the abnormalities typical of this syndrome, but the syndrome was named in honor of Alfred Poland in 1962 in a work by Clarkson (cited in Shamberger, 1998; Slezak and Sasiadek, 2000).

Etiology and pathogenesis

The etiology and pathogenesis of the disease are still unknown. The generally accepted theory for the development of the Poland syndrome is a misperfusion along the subclavian and/or vertebral arteries and their branches at week 6 of intrauterine development. The severity of the syndrome manifestations is determined by the duration and intensity of the circulation failure of these arteries [5]. Most cases of the Poland syndrome are sporadic, with only rare hereditary factor (1%). This syndrome is considered to have an autosomal dominant type of inheritance [6].

Czeizel et al. (1990) examined 18 typical patients with Poland syndrome and only one patient was burdened with a hereditary anamnesis. In the father, the greater pectoral muscle was absent on the left with severe hypoplasia of the thumb of the left hand and syndaktylia of the II and III fingers, whereas the son had moderate hypoplasia of the greater pectoral muscle on the left with severe hypoplasia of the thumb of the left hand [7].

In 2014, Vaccari et al. reported a case of Poland syndrome in monozygotic twins with a deletion of 11q12.3 involving five genes, four of which (*HRASLS5, RARRES3, HRASLS2*, and *PLA2G16*) are responsible for the regulation of cell growth, cell differentiation, and apoptosis via the ras-signaling pathway. Abnormality of these mechanisms of development can lead to oncological diseases, in addition to the abnormality of the differentiation of muscle cells [8].

In 1977, McGillivray and Lowry published the results of an examination of 44 patients with this syndrome, whose family history was not burdened. All the patients had no sternal part of the greater pectoral muscle on one side, with 40 of them having developmental disorders of the upper extremity. Only four of the patients had an intact upper extremity [9].

In the literature, some diseases manifest as unilateral hypoplasia of the gluteus muscle with foot symbrachydactylia, which are formed in utero, likely due to misperfusion along the external iliac artery, similar to the clavicular artery in Poland syndrome. By the opinion of several authors, these diseases may be similar conditions to Poland syndrome except for the lower extremity [10]. Riccardi (1978) and Corona-Rivera (1997) each described one case of unilateral hypoplasia of the dorsal gluteal muscle with foot symbrachydactylia [10, 11]. In 1995, Parano et al. published a report on a family in which three people (all women) had unilateral hypoplasia of the dorsal gluteal muscle, and their common ancestor had a unilateral aplasia of the greater pectoral muscle [12]. Since 1841, approximately 500 cases of Poland syndrome have been described worldwide.

Clinical characterization

The most detailed clinical characterization of Poland syndrome was found in the work by Yiyit

et al. (2015), based on the results of examinations of 113 patients aged 6–38 years. The authors reported a right-handed lesion in 55.7% cases, a left-handed lesion in 37.1% cases, and a bilateral lesion in 7% of observations. According to Jones (1926) and Lord et al. (1990), a right-sided lesion occurs in 75% of cases [cited in: Jones (1926); Lord et al. (1990)].

The main clinical sign of Poland syndrome is a lesion of the greater pectoral muscle. According to Yiyit et al. (2015), aplasia of the entire greater pectoral muscle was noted in 71.6% of 113 patients, while the sternocostal region had aplasia in 28.3% of cases. In addition to the greater pectoral muscle, other muscle lesions occur: the smaller pectoral muscle, the serratus anterior, the rhomboid muscle, the trapezius muscle, the abdominal rectus muscle, and the broadest muscle of back [2].

David and Winter (1985) reported a family in which men for three generations had unilateral aplasia of the large pectoralis muscle, the serratus anterior, and the broadest muscle of back. The authors emphasized that aplasia of the other muscles of the shoulder girdle, in addition to the greater pectoral muscle, can be crucial for choosing surgical treatment tactics; in particular, the inability to use the broadest muscle of back for reconstruction of the greater pectoral muscle [13]. In most patients with Poland syndrome, the chest has a unilateral deformity due to hypoplasia or aplasia of the costal cartilage (more often II–IV or III–V ribs) [14].

Yiyit et al. (2015) revealed absence of anterior part of the ribs in 20.3% of patients, and 7.9% had rib hypoplasia. The most frequent abnormality of the chest was a keeled chest (9.7%), followed by a hollowed chest (0.8%). A defect of more than one rib that led to the formation of pleurocele occurs in 7% of patients, while 4.4% of patients report paradoxical respiration [2]. Lieber et al. (2012) described the presence of the anterior part of the V rib, the hypoplasia VI, and the concrescence of VII and VIII ribs on the affected side of the defect [15].

Yiyit et al. (2015) revealed scoliosis in 1.7% of cases. In patients with left-sided manifestation of Poland syndrome, Yiyit et al. (2015) found the complete manifestation of *situs inversus* in no cases (in 7% of cases only dextrocardia was detected). In more than half of the patients with dextrocardia, there was a partial absence of ribs on the side of the lesion. No heart defects were diagnosed [2].

The overwhelming majority of patients had thinning of the subcutaneous fat on the side of the lesion (86.7%). The characteristic symptoms of deficiency of hair on the chest (59.2%) and axillary cavity (64.6%) on the side of the lesion were noted [2]. In most cases, patients with Poland syndrome had abnormalities of development of the breast and the nipple-areolar complex on the side of the lesion [displacement upwards (57.5%), hypomastia (64.6%), hypotelia (61%), amastia and atelia (2.6%), and polytelia (0.8%)] [2]. An evaluation of the severity of the chest deformity is difficult due to the polymorphism of the clinical manifestations of Poland syndrome.

Seyfer et al. (2010) defined two forms of the syndrome depending on the nature of the chest lesion: simple (there is only a defect of soft tissues) and complex (defect of soft tissues and the skeleton of the chest) [16]. Currently, in clinical practice, there is a common classification of Foucras with the definition with three degrees of severity [1].

Degree 1: minor deformity due to hypoplasia of the greater pectoral muscle and moderate hypoplasia of the mammary gland, leading to asymmetry in the mammary glands in women and barely noticeable asymmetry in the chest of men. The nipple-areola complex is smaller in size compared to the healthy side, and it is located higher. There are no skeletal abnormalities.

Degree 2: moderate deformity with noticeable aplasia of the greater pectoral muscle, hypoplasia of other pectoral muscles (serratus anterior, external oblique muscle, and the broadest muscle of back), moderate deformity of the ribs, and marked deformity of the chest in both women and men. There is severe hypoplasia or complete aplasia of breast tissue with the nipple-areolar complex being hypoplastic or completely absent.

Degree 3: severe chest deformity due to aplasia of the breast, greater pectoral muscle, and other muscles of the chest. There is aplasia of the ribs and deformity of the sternum.

Congenital malformations of the hand are characteristic of complete Poland syndrome, and are identified on the same side as the lesion of the chest. The incidence rate of hand damage is 12–56%, according to different authors [1, 2]. The severity of their manifestation is different; some patients have shortening of the middle phalanges of the fingers in combination with syndactylia, while some have complete absence of the fingers of the

hand. In 1989, Shamberger et al. reported that 67% of the studied patients with Poland syndrome had unilateral syndactylia with the most common lesion of the II–IV fingers [17, 18]. There are various classifications of congenital malformations of the upper extremity in this syndrome, but the most complete is the classification of Al-Qattan (2001), which includes the definition of seven types.

Type 1—normal hand, there is only a deformity of the chest.

Type 2—deformity of a hand is insignificant and manifested only when compared to the contralateral side.

Type 3—the classical form (symbrachydactylia).

Type 4—there are several functioning rays on the hand: A—radial clubhand with a dangling thumb or aplasia of the first ray; B—adactylia of the II finger; C—adactylia of the II-III fingers; D—adactylia of the central rays (cleavage of the hand); E—adactylia of the ulnar fingers.

Type 5—all fingers are not functioning or missing.

Type 6—transverse defects proximal to the metacarpophalangeal joints.

Type 7—deformity of the upper extremity, similar to phocomelia [19].

In addition to the abnormalities of the musculoskeletal system in Poland syndrome, it can involve other organs and systems in the pathological process. For example, cases of damage to the hematopoietic system are described [20], such as leukemia [21] and non-Hodgkin's lymphoma [22]. In 2006, Jacob Ndas Legbo described a case of a 12-year-old girl with Poland syndrome and severe thrombocytopenia. In the anamnesis, the patient had frequent hemorrhages in the area of the anterior thoracic wall on the side of the lesion. It was not possible for the author to explain such selectivity of the lesion [23]. For orthopedic pathologies, Yiyit et al. (2015) observed Sprengel disease in 15.9% of patients [2].

Treatment

The tactics for treating patients with Poland syndrome is determined by the form of pathology (complete and incomplete), the severity of anatomical and functional changes, and by age.

We can distinguish two main groups of surgical interventions.

1. Reconstructive interventions on the hand (upper extremity).
2. Reconstructive interventions on the chest.

Reconstructive interventions on the hand (upper extremity)

In patients with the complete form of Poland syndrome, the surgical treatment starts with restoring the function of the hand grasp and improving the possibility of self-service. The main variants of hand interventions are elimination of the syndaktylia, restoration of the opposition of the first ray, extension of the fingers. Transplantation of the toes to the hand is performed in extreme forms of underdevelopment [24, 25].

Reconstructive interventions on the chest

The choice of treatment tactics for patients with Poland syndrome with chest deformities is determined by the nature and severity of anatomical and functional disorders, age, gender, and the patient's desire. If the defect is limited to soft tissues, then the reconstructive interventions are performed for aesthetic indications. If the patient has defects in the ribs or sternum or paradoxical respiration, the reconstructive interventions aimed at restoring the thoracic cage are required to improve lung function, to prevent the progression of respiratory failure, and to protect the mediastinal organs [26–28]. For aesthetics, women with Poland syndrome need to restore the volume of the breast and nipple-areolar complex, fill the soft tissue depression in the subclavian area, and restore the symmetry of the chest. In men, the purpose of the surgery is to increase the volume of the soft tissues of the chest, to mask the ribs, and to restore the symmetry of the chest [29].

There are various opinions on the timing to start treatment of patients with Poland syndrome. Some authors believe that surgery should be performed at the end of adolescence, which will enable maximum symmetry and prevent subsequent interventions [2, 29], while others show good results in treatment of children at an earlier age [30, 31].

The methods of correction of chest deformities can be divided into the following groups:

- restoration of the contours of the chest (autografting of skin-fat and muscle grafts,

lipografting, injection of polymer polyacrylamide solution, prosthetics);

• reconstruction of the breast (autografting of skin-fat and muscle grafts, prosthetics, lipografting, formation of a nipple-areolar complex);

• restoration of the frame function of the chest (restoration of rib defects, elimination of pleurocele, as well as deformities of the sternum).

To date, it is preferred to transplant the broadest muscle of back (in free and non-free versions) to the position of the greater pectoral muscle in the reconstruction of chest deformities in patients with Poland syndrome worldwide [3]. In the opinion of Popodopulos et al. (2011), the broadest muscle of back enables one to fill the necessary volume of soft tissues of the chest, similar to the greater pectoral muscle, and gives the best results of treatment [3]. Transposition of the broadest muscle of back enables not only to restore the contours, but also to stabilize the chest, and provide additional coverage of the prosthesis of the gland (when it is used), as well as increases its stability [29]. In case of a deficiency of soft tissues on the anterior surface of the chest, the first stage is an expander dermotension, followed by grafts transplantation if necessary, in conjunction with the installation of a breast implant. In the absence of the broadest muscle on the side of the lesion, it can be transplanted from the contralateral side [32].

Popodopulos et al. (2011) assessed the quality of life of 49 women with Poland syndrome who underwent various interventions on the chest: 16 underwent the transposition of the broadest muscle of back, 12 underwent expander dermotension followed by installation of the breast implant, and 4 underwent the transposition of the transverse rectus abdominis myocutaneous (TRAM) flap (flap based on the abdominal rectus muscle). Sixteen patients were satisfied with the results (13 patients underwent transposition of the broadest muscle of back, 2 of them underwent installation of a breast implant, and 1 underwent transposition of the TRAM flap).

In the literature, single cases of the use of free autografts for the chest and breast reconstruction in patients with Poland syndrome are described (lower-gluteal and upper-gluteal flaps, a flap based on the superficial lower epigastric artery, a deep inferior epigastric perforators flap (perforating flap based on the lower epigastric artery), an epiploon, an anterolateral flap of the thigh, and a musculocutaneous flap based on *m. gracilis* [33–37]. The disadvantage of these interventions is a high percentage of complications. The advantage is the minor damage to the donor area, as well as the rapid recovery of patients compared to the transposition of the broadest muscle of the back [38].

An alternative method of aesthetic correction of chest deformity in Poland syndrome is the use of silicone implants. Fekih et al. (2010) used this technique in the treatment of deformities of II–III degrees [39, 40]. Soccorso et al. (2015) reported on the experience of treating a hollowed chest in a child with Poland syndrome with a silicone implant, and the reported a good cosmetic result [31]. According to Seyfer et al. (2010), the use of silicone implants has a high percentage of complications [16] and in complex forms of the syndrome, the implant installation is extremely difficult [36].

One of the methods of cosmetic correction of the chest deformity is microinjection autografting of fatty tissue or lipografting. This is a procedure in which the fatty tissue of the patient is taken from the hips, abdomen, and breech by means of liposuction and injected into the area where it is necessary. The main advantages of this method of correction are absence of a rejection reaction, low invasiveness, and a minimal rehabilitation period [41]. Due to resorption of the fatty tissue, in order to obtain a satisfactory aesthetic result, multiple interventions are required [38]. Pinsolle et al. (2008) reported using this technique in 8 patients (7 women and 1 man). The number of injections was from 1 to 5 and the average volume was 96 ml (from 25 to 200). In one case this method was used in an isolated form, and in seven cases, it was used in combination with reconstructive surgeries. According to the authors, lipografting can be used to treat patients with chest deformities in Poland syndrome as an independent procedure, but the best results are found when it is combined with other methods. Injections of adipose tissue enable a correction of the defects of the contours of the chest [32]. Fekih et al. (2010) described the experience of performing liposuction from the contralateral side to eliminate chest asymmetry in patients with moderate deformities. In some cases, in order to maximize the correction of the shape and size of the breast, reduction interventions from the contralateral side are required [39]. He et al.

(2016) described the technique of a one-stage nipple repair using a *thoracodorsal artery perforator* (TAP) flap (skin-adipose) and the gland itself, forming a muscle flap based on the broadest muscle of the back in patients with Poland syndrome. During the surgery, a tissue expander was installed under the muscle with dermotension started a week later. After 2 months, the second stage was performed, which was removal of the expander and installation of an implant of the required size. This technique was used in 12 female patients aged 15–21 years, with good and very good results noted in 11 cases [42].

In those cases when patients have severe abnormalities in the development of the sternum and ribs that cause functional or gross cosmetic disorders, reconstructive surgeries on the thoracic cage are initially performed. These include: a Ravich's thoracoplasty and its modifications [29]; replacement of defects with cartilaginous autografts (costal cartilage is taken from the opposite side), bone-cartilaginous allografts, artificial materials, titanium mesh [15]; Nuss modified thoracoplasty [30]; osteoplastic surgeries, including a one-stage osteotomy of the sternum and transposition of ribs with a vertical expandable prosthetic titanium rib fixation [15]; and replacement of the rib defect along the anterior surface of the chest with titanium plates using the Matrix rib system [26].

Seyfer et al. (2010), analyzed the long-term results of treatment of 63 patients with Poland syndrome with a follow-up of 1–21 years after the surgery and concluded that in simple forms of the syndrome, the transposition of the broadest muscle of the back to the position of the greater pectoral muscle in men, and the addition of this intervention in women with the installation of a breast implant under the transposed muscle is indicated. In complex cases, in addition to transposition of the broadest muscle of the back, it is also necessary to restore the skeletal function of the chest [16]. Foucras et al. (2003) follow a similar algorithm for treatment. The authors point out the advisability of using lipografting as an additional procedure to improve the symmetry of the chest [1].

Conclusion

The presented review of the literature highlights comprehensively the issues of etiopathogenesis, clinical treatment, modern technologies for rehabilitation of patients with Poland syndrome, and reveals the advantages and disadvantages of the existing methods. Given the rare frequency rate, the complexity of pathology, and the polymorphism of clinical manifestations, and the lack of a unified approach to treatment, this topic requires further study and more research investigations.

Information on funding and conflict of interest

The work was performed with the support of the Turner Scientific and Research Institute for Children's orthopedics, the Ministry of Health of Russia. The authors declare no obvious and potential conflicts of interest related to the publication of this article.

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