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Chest wall reconstruction in Poland Syndrome: An analysis of surgical techniques and clinical manifestations

Pedro Fechine Honorato, Anna Vitória Paz Moreira, Dhiego Alves de Lacerda, Isabelle Lima Lustosa, Renata Silva Cezar, Arthur de Sousa Lima Carvalho, Jalles Dantas de Lucena[1].

INTRODUCTION

Poland Syndrome, identified in 1841, is a rare congenital malformation characterized by the absence of the sternocostal portion of the pectoralis major muscle, manifesting unilaterally and presenting symptoms such as thoracic asymmetry and breast hypotrophy (URSCHEL, 2009; MASIA et al., 2015). In severe forms, there are bone and kidney alterations, and it is recognized as a genetic condition with an incidence ranging from 1:7,000 to 1:100,000 live births (DELAY; NACHAOUI; FROBERT; 2022; KIZILYEL et al., 2023).

Poland Syndrome is notable for the absence of pectoral muscles, breast hypoplasia, and rib defects, with a multifactorial etiology involving genetic and environmental factors, possibly related to subclavian artery involution at 6 weeks of gestation (STYLIANOS et al., 2011). The diagnosis, often underdiagnosed due to the aesthetic focus, is usually clinical, with imaging and aspiration cytology to confirm the diagnosis (HUANG et al., 2018; LYMPEROPOULOS; RAMADAN; KOSHY, 2020).

Several surgical techniques are used to correct the syndrome, such as myocutaneous flaps, silicone implants and fixation of metal bars, adapting to the specific needs of the patient (LYMPEROPOULOS; RAMADAN; KOSHY, 2020). Thus, the present study aims to describe the main clinical findings of Poland Syndrome and surgical techniques available for correction.

MATERIALS AND METHODS

This study is a literature review, carried out in February 2024 in the online databases: Virtual Health Library Portal (VHL), *PubMed, SciELO, Medline* and *Google Scholar*. For the search for studies, descriptors related to the theme were selected in the Health Science Descriptors (DeCS): "Anatomy", "Surgery" and "Poland Syndrome", in addition to equivalent terms in the Medical Subject Headings (MeSH): *"Anatomy", "Surgery"* and *"Poland Syndrome".* The search was performed using the "*AND" operator* for effective cross-referencing of the descriptors.

[1] Santa Maria University Center - UNIFSM, Cajazeiras – PB

The inclusion criteria used were to select full articles available online, free or paid, published in the period from 2007 to 2024, that reported cases in adult humans (19-44 years) and published in Portuguese, English, and Spanish. On the other hand, some exclusion criteria were established, excluding research carried out on animals, articles not aligned with the objectives of the study.

After the strict application of the inclusion and exclusion criteria, 15 relevant articles were identified and analyzed in full. It is important to highlight that this research is a literature review and did not involve the participation of human beings, thus dispensing with submission to the Research Ethics Committee (REC).

RESULTS

Poland Syndrome is a rare congenital malformation, presenting more frequently in males (MASIA et al., 2015). Characterized by the absence or hypoplasia of the pectoralis muscle, this syndrome can present severe manifestations, such as syndactyly, monodactyly, dextrocardia, breast hypoplasia, pulmonary hernia, genitourinary and vertebral anomalies (ORTIZ, 2014).

The syndrome involves hypoplasia or agenesis of the anterior part of the chest muscles, mainly affecting the sternocostal portion of the pectoralis major muscle, followed by the pectoralis minor muscle (ERGÜVEN et al., 2011). This complexity of manifestations, combined with its congenital origins, highlights the clinical diversity of Poland Syndrome, making a comprehensive approach to its diagnosis and management essential.

The pathogenesis of Poland remains poorly understood in the literature, with suggestions of a possible vascular insult during the sixth week of gestation, involving the division of the pectoral mass and vascular differentiation of the aortic arches (CINGEL et al., 2013). This syndrome is characterized by unilateral agenesis of the pectoral muscles, often accompanied by ipsilateral brachysyndactyly, presenting variations such as nipple involution, breast tissue, hypoplasia of the costal arches, and shortening of the ipsilateral upper limb (BAZZI JUNIOR et al., 2012).

In addition to musculoskeletal characteristics, the syndrome may be associated with several complications such as: cardiac, renal and other congenital abnormalities, with reports of coagulation disorders, psoriasis, lupus, leukemia and non-Hodgkin's lymphoma, among others (GASHEGU et al., 2009; LACUT; MARSEILLES; GUERRINI, 2010; RIBEIRO et al., 2009). The presence of dextrocardia occurs in 5% of cases, with a generally favorable prognosis, without association with other cardiac pathologies (JIMÉNEZ et al., 2009).

Diagnosis involves periodic laboratory and imaging tests, such as radiographs of the upper limbs and chest, echocardiogram, renal and abdominal ultrasound, blood count, and urine (BAZZI JUNIOR et al., 2012). The syndrome exhibits muscle and bone involvement, and can be classified as mild, moderate,

and severe, with different degrees of involvement of the pectoralis muscle, thoracic skeleton, and other anatomical structures (ORTIZ, 2014; DOLAS et al., 2014).

In its mildest presentation, the syndrome is characterized by hypoplasia of the pectoralis major muscle and breast, with elevation of the nipple-areolar complex. In the moderate form, the absence of the pectoralis muscle and hypoplasia of the breast, nipple and areolar complex are observed. Both forms do not compromise the thoracic skeleton. In the severe form, in addition to the above-mentioned alterations, there are deformities in the thoracic skeleton, involving the sternum and ribs (DOLAS et al., 2014).

The condition, because it is sporadic, presents a wide variation of symptoms and clinical manifestations, making diagnosis challenging in some cases. In addition, the syndrome is often underdiagnosed, especially when patients report only aesthetic problems. Seeking medical help is usually due to concern about appearance, especially among women during adolescence (ORTIZ, 2014).

In diagnostic screening, ongoing research is crucial to unravel the exact mechanisms that lead to the development of the syndrome, providing insights for early diagnosis, clinical management, and future therapeutic approaches (BAZZI JUNIOR et al., 2012; KASAR; ERTEKIN, 2022).

The therapeutic approach to Poland Syndrome is highly individualized, considering a variety of factors, such as the degree of the deformity, the patient's age, gender, and personal preferences. In cases where the malformation is limited to the pectoralis major muscle, especially in men, surgical intervention may not be necessary, often opting for therapeutic abstention (JIMÉNEZ et al., 2009).

Among the therapeutic options, the use of breast/pectoral prosthesis implants is a common choice, with the insertion of breast implants in women and pectoral implants in men, each through a specific axillary approach. These procedures are not without risks, such as hematoma formation, infection, implant migration, and capsular contracture (FEKIH et al., 2010). Reconstruction of the pectoralis muscle can be achieved by means of ipsilateral latissimus dorsi muscle flaps or the rectus abdominis cutaneous muscle flap (TRAM), although the latter is contraindicated in young patients to avoid complications such as abdominal wall weakness (NISHIBAYASHI et al., 2013; DOLAS et al., 2014).

Lipomodeling, using an autologous adipose tissue graft, is another alternative, using common donor areas, such as the abdominal and thigh regions. This technique offers significant modeling capacity, giving the breast a more natural and symmetrical appearance (MASIA et al., 2015). Buchanan et al. (2016) highlighted functional muscle transfer as an option, emphasizing the importance of patient motivation to follow an exercise program aimed at improving upper extremity strength and function.

Several surgical techniques have been proposed to correct the deformities associated with Poland Syndrome, and the choice between them depends on the specific characteristics of each patient (BORSCHEL; COSTANTINO; CEDERNA, 2007; RESENDE; FAN; CRUZ, 2011; BUCHANAN; LEYNGOLD; MAST, 2016; TOMÁS et al., 2013). In situations of large defects, surgical intervention

after the growth spurt, performed by a team experienced in pediatric thoracic surgery and plastic surgery, is often recommended. Deep defect repair seeks to provide a stable basis for implants and tissue transfers, and is indicated for cases with more significant deformities of the chest wall and overlying soft tissues. The decision to proceed with deep reconstruction after puberty is highly dependent on the patient and surgical experience. More superficial defects can be corrected directly, without the need for a base coat of prosthetic tissue (MOIR; JOHNSON., 2008).

FINAL THOUGHTS

Poland Syndrome exhibits diverse clinical manifestations, from hypoplasia of the pectoralis muscle to complications such as dextrocardia. Diagnosis, often clinical, requires imaging tests for comprehensive evaluation, including x-rays and echocardiogram.

In the surgical approach to the syndrome, several options are considered, such as breast/pectoral prosthesis implants, muscle flaps, lipomodeling, and functional muscle transfer. Individualization in the choice of technique is essential, adapting to the specific needs of each patient and the extent of the deformity, emphasizing the importance of a personalized approach in the treatment of Poland's multifaceted syndrome.

Keywords: Anatomy, Surgery, Poland Syndrome.

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