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Case Report

Poland's Syndrome Treatment with Customized Implant - A Case Report

Ricardo Cavalcanti Ribeiro*, Pedro Felipe Suárez and Avelino Monteiro De Abreu

Division of Plastic and Reconstructive Surgery, Carlos Chagas Institute, Brazil

***Corresponding author**

Ricardo Cavalcanti Ribeiro, Division of Plastic and Reconstructive Surgery, Carlos Chagas Institute, Hospital Casa de Portugal, Av Fernando Mattos, 225–Barra Da Tijuca–22621090 Rio de Janeiro, Brazil, Tel: 55 21 999861120; Email: rribeiro@centroin.com.br

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Keywords

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- Regular implant
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Abstract

Background: The Poland's syndrome is a rare congenital alteration of the thoracic region, mainly characterized by the partial or complete absence of the pectoralis major muscle. It has one higher incidence in males, with a predominance of the right side of the chest. Several factors have been associated as possible causes, but currently a main ethology has not been found.

Patient and methods: We report the case of a 32-year-old male patient, who presents absence of the pectoralis major on the right side, with only preservation of the portion upper distal of the muscle. Through the use of a mold that is adapted to the region of the defect, we obtained a customized implant with exact measurements of the desired area, the which was placed the day of surgery. A 3D thoracic computerized tomography was perfomed in order to achieve better shape.

Results: With the use of this customized prosthesis, we achieve adequate correction of the defect, obtaining the desired symmetry of the thoracic region and a rapid recovery of the patient, without any complications during surgery.

Conclusion: Using this approach best results are obtained, with less morbidity for the patient, fewer adverse effects and a more rapid recovery, making it an excellent choice in the treatment of sequelae of Poland's syndrome.

INTRODUCTION

Poland's Syndrome (PS) is a rare abnormality, with a sporadic presentation, characterized by congenital malformations of the chest wall, with or without alterations to the ipsilateral superior limbs and hands [1,2]. Classically, it consists of a combination of unilateral aplasia of the sternocostal portion of the pectoralis major muscle (PMM) and hypoplasia of the ipsilateral hand, with syndactyly [3,4] and synbrachydactyly [5]. Over 400 cases of PS were reported as of 1990; in these cases, several degrees of chest wall malformations extending to the superior limbs were seen [1]. The incidence of PS has a male: female ratio of 3:1. According to the literature, it is a rare disease, with a frequency estimated at 1 in 30,000 live births [6,7] with the right side affected twice as often as the left side [8,9]. The etiology of PS is still unknown.

Multiple therapeutic approaches have been made in order to correct this abnormality of the thorax [10-11]. Among the techniques of correction in male patients we have reconstruction with latissimus dorsi myocutaneous flap, use of the transverse abdominal flap, tissue expander, polymeric polyalkylimide solution injection, customized titanium implant for chest wall reconstruction [12], until simple placement of fat graft with the aim of improving the appearance in this region [13].

Our goal is to present a fast and effective therapeutic alternative, with low rate of complications and rapid recovery for a patient with Poland's syndrome, through the use of a customized implant.

CASE REPORT

Male patient 32-year-old, with obvious asymmetry in the chest on the right side, present from birth. No account of Comorbidities, with negative family history for deformities and genetic alterations. Patient concerns a good psychomotor development without alteration of the force. As compensation, it presents a hypertrophy of the adjacent muscles associated with a great sports activity throughout its development.

To the physical and radiology examination, helped with the 3D thoracic computerized tomography, was noted an almost total absence of the pectoralis major muscle, with only preservation of the distal portion that is inserted into the inferior border of

the clavicle, with little alteration in the axillary region on the same side (Figure 1). Other associated malformations are not readily apparent. Functionally, the patient has excellent mobility of upper limbs, without limitation for adduction, abduction and elevation of the arm ipsilateral to the aforementioned deformity.

For the resolution of this pathology, a therapeutic option is presented for the patient, through the placement of customized implant, with prior use of a mold that fits the area to rebuild. This mold was sent to a company (Polytech Health & Aesthetics®. Dieburg–Germany) in order of the elaboration of such implants with exact measurements of the defect (Figure 2).

SURGICAL TECHNIQUE

Patient underwent to general anesthesia in dorsal decubitus, infiltrates the right thoracic region with anesthetic solution more epinephrine. A lateral incision is performed, taking advantage of the area tattooed in order to hide the scar. Is performed the adaptation of the prostheses made by measure in the right chest region, resulting in an immediate satisfactory result. As a preventive measure, a vacuum drain was placed to reduced fluid colections, until drainage inferior than 35 ml (Figure 3-4).

DISCUSSION

Poland's syndrome is consider a congenital malformation of the thoracic region, characterized by absence of the of the pectoralis major muscle. As a result, evidenced a lack of anterior axillary pocket.

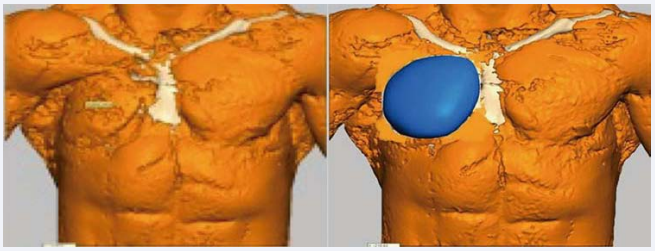

Figure 1 3D thoracic computerized tomography. Is performed to the patient in their pre and postoperative period, in which there is evidence of an adequate coverage of the defect, obtaining the Symmetrization of the thoracic region.

Figure 2 It is performed the full coverage of the defect in right thoracic region, through the application of a mold that is used for the manufacture of a customized prosthesis. This prosthesis is placed in the surgery in order to correct this imbalance. The mold is sent to a company (Polytech Health & Aesthetics®. Dieburg – Germany) that is in charge of the preparation of the implant.

Figure 3 It is observed the customized implant, occupying the place of pectoralis major muscle absent. This type of implant allows full coverage of the defect with with the thickness, size and format required for adequate correction of the defect.

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It has an incidence of 1 in 10,000 to 1 in 100,000 live births. Usually in female patients her diagnosis is made only after the breast development and referred by the patient as an asymmetry in the mammary region. In male patients, the suspect diagnosed is made by clinical examination, with obvious asymmetry in the thorax, posterior to the full development of this region. Their form of presentation is 3:1 more frequent in men than in women, with a predominance of the right side in the male patient, as it was observed in our case and reported in the literature.

With the emerging of new technology, the correction of the deformity is made simpler and with more satisfactory results. As an alternative resource we have the development of an customized implant that allows for accurate coverage of the defect. Currently exists in the market a wide range of implants not customized, with the lowest values, but in return do not allow a precise modeling to the defect, since they have standard measures that are not adapted fully to the desired area.

Through the previous elaboration of a mold that is adapted to the desired location, you get an accurate measurement of the area to be treated. With the help of imaging studies, is performed a prosthesis with the thickness, size and format required for

adequate correction of the defect. This resource presents less morbidity to the patient, with faster recovery and a complication rate lower when compared to other types of procedures.

CONCLUSION

The Poland syndrome is a rare disease of variable presentation, with greater predominance in male patients. For the plastic surgeon, it is of great importance the treatment of these patients in order to improve their physical appearance, functional as its psychosocial state, using an approach that allows the appropriate resolution of this defect.

With the implementation of new technologies as the customized implant, better results with less morbidity were achieved, as well, fewer adverse effects and a faster recovery for the patient, being an excellent choice for the correction of the defect caused by the Poland´s Syndrome, as demonstrated in this case.

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