Poland 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.\(^5\) Only 400 cases of PS were reported by 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-to-female ratio of 3:1 and its frequency is estimated at one in 30,000 live births,\(^6-11\) with the right side being affected twice as often as the left side.\(^12,13\) The etiology of PS is still unknown.

Several studies have suggested that a genetic factor or, even more likely, extrinsic factors between the sixth and eighth weeks of pregnancy may interfere with PMM migration and the separation of the digits (chirodactile) that occurs in this period.\(^6-12\) The recent vascular hypothesis for the etiology of PS implies that hypoplasia of the ipsilateral subclavian artery is the origin of this birth anomaly.\(^13-16\)

The main deformities associated with PS are as follows:

1. Muscles—Absence of the pectoralis minor muscle (PMM), absence of the clavicular and sternal segments of the PMM,\(^17\) and hypoplasia of the serratus anterior,\(^18,19\) latissimus dorsi, deltoid, and infraspinatus and supraspinatus muscles.\(^20\)

2. Mammary glands—Areolar abnormalities, mammary absences, or hypoplasia.\(^17,18,21\)
The risk of lung herniation during respiration. Unilateral PS4–29 and are not uncommon conditions in women seeking mammary augmentation for breast asymmetry.13

Clinical and radiographic Poland syndrome classification

<table>
<thead>
<tr>
<th>Degree of presentation</th>
<th>Mammary alterations</th>
<th>Musculoskeletal chest alterations</th>
<th>Ipsilateral superior limb alterations</th>
<th>Other congenital alterations</th>
</tr>
</thead>
<tbody>
<tr>
<td>First (mild)</td>
<td>Amastia; hypomastia or areolar asymmetry</td>
<td>None, or partial absence of pectoralis major muscle</td>
<td>None</td>
<td>May be present</td>
</tr>
<tr>
<td>Second (severe)</td>
<td>Hypomastia or amastia; areolar asymmetry</td>
<td>Total absence of pectoralis major muscle; different alterations of the muscles and/or bones of the ipsilateral chest</td>
<td>No or small alterations</td>
<td>May be present</td>
</tr>
<tr>
<td>Third (very severe)</td>
<td>Amastia; areolar asymmetry</td>
<td>Different manifestations</td>
<td>Present</td>
<td>May be present</td>
</tr>
</tbody>
</table>

(3) Thorax—Pulmonary herniation, hypoplasia of the ribs, or deformities of the hemithorax, depending on severity.22

(4) Ipsilateral superior limb—Skeletal hypoplasia affecting the hands,19 forearms, upper arms, and scapula; syndactyly, symbrachydactyly,17 polydactyly, brachysyndactyly, and axillary constrictions.3–23

Other abnormalities associated with PS include dextrocardia, kidney hypoplasia,18–24 neurofibromatosis,9 myasthenia gravis,8 peroneal atrophy (Charcot-Marie-Tooth Disease), microcephaly, craniofacial dysplasias, and Möbius syndrome.25,26 There have also been reports of associated coagulation disturbances, psoriasis, systemic lupus erythematosus, Baselow–Graves disease,27–29 leukemia, and non-Hodgkin lymphoma.30 All of these components are not necessarily always present, but the full syndrome consists of an assortment of these abnormalities.14

PS is characterized by different degrees of mammary hypoplasia affecting the subcutaneous tissue and the adjacent muscles, moreso when costal arc deformities and/or syndactyly are also present.22 The spectrum of the deformities varies and may involve the simple absence of the sternocostal segment of the PMM or the total absence of all the chest wall components except for the skin and the pleural membranes,22 a condition that is associated with the risk of lung herniation during respiration.31 Unilateral hypoplasia of the breast and PMM without upper limb involvement are seen more often than other forms of PS4–29 and are not uncommon conditions in women seeking mammary augmentation for breast asymmetry.13

Imaging techniques have a fundamental role in the detection, location, and characterization of chest wall disorders, many of which have characteristic radiologic appearances that allow definitive diagnosis. Sternal deformities can be seen with radiographic examinations and their severity quantified with computed tomography (CT) scans,32 which clearly show many abnormalities.31 For chest wall disorders, the chest radiograph remains the basic imaging study for evaluating pathology. Chest radiographs generally show a hyperlucency on the affected side, which mimics a radical mastectomy.22–29,31 A mammogram is useful in showing breast asymmetry or the absence of the PMM. PS has been diagnosed in one of 19,000 mammograms,33 according to Perez Aznar et al.34 Failure to show the PMM on a mammogram is most often related to errors in positioning. Ultrasoundography also has a complementary role in diagnosing thoracic pathology.34

The advent of cross-sectional imaging techniques, such as CT and magnetic resonance imaging (MRI), has enabled the precise localization of chest wall lesions and, in some cases, definitive diagnoses.32 The MRI shows a better tissue contrast, but is less useful than CT in the study of bone pathologies.25 CT also depicts more clearly the absence of the PMM and allows for the better appreciation of nearby associated musculoskeletal anomalies,32 such as mediastinal widening.35 CT and MRI together define, with precision, the musculoskeletal abnormalities, making these tools useful in the presurgical period. Hurwitz et al.22 justify the use of three-dimensional (3-D) CT among all available imaging studies for the planning of surgical procedures to assure the best possible reconstruction. This imaging technique can offer an accurate representation of tissue deficits and asymmetry, thereby helping clinicians make decisions regarding the transposition of muscular flaps and reconstructive techniques.

In the current retrospective study, 28 female patients were evaluated over a 17-year period. Male patients were excluded because of differences in mammary gland development and to facilitate a standard classification and treatment algorithm. Patients had different levels of thoracic and superior limb compromise.

Based on the analysis of the imaging examinations and a clinical evaluation, the cases were divided into groups based on a new classification system. This proposed clinical and radiographic PS (CRPS) classification was designed to facilitate an accurate and practical study of female patients and to assist in choosing the best surgical approach. An algorithm for the planning of surgical treatment is also presented.

METHODS

Data were obtained from the 28 female patients with PS who were treated in the 28th Infirmary, Plastic and Reconstructive Surgery Division of the Hospital Santa Casa da Misericórdia do Rio de Janeiro. Imaging studies were performed by the Radiology Department of the Universidade Federal do Rio de Janeiro between February 1992 and January 2009. For each patient, we determined the degree of severity and absence or presence of congenital anomalies (Table 1). Only female patients were included to standardize the thoracic findings in mammary...
gland development, which is a variable of the CRPS system. The degrees of severity are listed below.

First Degree (Mild). The diagnosis of first-degree PS would be made in a patient with mammary asymmetry caused by hypomastia or amastia and areolar asymmetry, with or without a partial absence of the PMM. No other musculoskeletal alterations are observed; other congenital alterations may or may not be present.

Second Degree (Severe). Hypomastia or amastia, areolar asymmetry, total absence of the PMM, and alterations of the ipsilateral muscle group and/or bones of the chest results in a diagnosis of second-degree PS; ipsilateral superior limb alteration and other congenital alterations may or may not be present.

Third Degree (Very Severe). Third-degree PS would be diagnosed in patients with amastia; areolar asymmetry; major ipsilateral musculoskeletal chest alterations, such as total absence of the PMM, the pectoralis minor muscle, and/or the serratus anterior muscle; possible lung herniation; widened opening of the mediastinum; and ipsilateral superior limb alteration. Other congenital alterations may or may not be present.

Imaging Studying Evaluation
All patients underwent chest radiographs and conventional thoracic CT scans; MRI was performed for patients 14 through 28 once this imaging modality became available in Rio de Janeiro. Three-dimensional reconstructions of the shape and volume of the cutaneous tissue, bones, and internal organs were conducted for patients 2, 3, 6, and 18. In addition, a helical thoracic CT was performed in patients 1, 8, 10, 15, 16, and 17 (Figure 1).

Figure 1. A, Roentgenographic study of a 32-year-old woman with deformities in the third to fifth ribs, mediastinal right deviation, and absence of the left breast silhouette. B, Axial computed tomographic images with contrast from a 10-year-old female, showing asymmetry in the chest silhouette in the frontal view, absence of the pectoralis major muscle, and hypoplasia of the pectoralis minor muscle. C, Magnetic resonance imaging scan with 3-D reconstruction shows chest asymmetry, right-sided thoracic deformities, mammary hypoplasia, absence of the pectoralis major muscle, sternum rotation, and mediastinal widening. D, Magnetic resonance imaging scan with 3-D reconstruction showing the same patient in part C after her first-stage procedure with subglandular placement of a tissue expander (round, 400 cc).
Surgical Technique
An algorithm for the planning of the surgical treatment for all 28 patients was created from the proposed CRS classification (Table 2). A total of 55 procedures were planned for surgical correction of aesthetic and/or functional abnormalities. Such corrections included the placement of breast implants, creation of customized breast implants, use of tissue expanders, and performance of local flap as well as latissimus dorsi flap surgery, the Ravitch procedure, and contralateral symmetrization procedures such as mammary reductions or augmentations. All patients with second- and third-degree PS were referred to thoracic and orthopedic specialists for evaluation before...
undergoing surgery. Different surgical techniques were used according to the degree of PS as shown below.

1. First degree (mild)—Breast implantation or customized breast implantation and contralateral mammary reduction or augmentation when needed (symmetrization procedure).
2. Second degree (severe)—Tissue expander placement when needed; regional local flap surgery; breast implantation or customized breast implantation; symmetrization procedure.
3. Third degree (very severe)—Tissue expander placement; latissimus dorsi flap or other flap surgery, such as a free flap or transverse rectus abdominis myocutaneous (TRAM) flap; breast implantation or customized breast implantation as needed; other surgeries such as the Ravitch procedure and a contralateral symmetrization procedure.

**RESULTS**

Twenty-eight female patients between 12 and 53 years of age presented with PS of different degrees of severity, with or without other congenital anomalies. The data for each patient contained a synopsis of the anomalies, which included demographic information, family history, concomitant morphologic or other alterations, and the information obtained from the imaging studies (Table 3).

Ten patients had first-degree PS, 14 patients had second-degree PS, and four patients had third-degree PS. Table 4 shows the CRPS level of classification for each case and the distribution of these classifications. Twenty patients had their right side affected; eight were affected on their left side.

All surgical corrections were based upon the severity of disease according to the radiographic images. Ten patients had not yet undergone surgery at the time that this report was written: five were undergoing preoperative screening and five were too young for surgical resolution because their growth was not yet complete. Eighteen patients underwent thoracic surgical procedures for reconstruction; the types of procedures are summarized in Table 5.

Eight patients with first-degree PS underwent surgical corrections, with seven receiving breast implants placed in a submuscular plane and one receiving a customized breast implant placed similarly; three of them required reduction of the contralateral breast for symmetrization (Figure 2).

Among the seven second-degree PS patients who underwent surgery, three received tissue expanders as an initial approach to treatment. After implantation, the volume of the prosthesis was expanded each week under general anesthesia until the maximal expansion was achieved. This process required 90 postoperative days. Two of these patients underwent a second procedure involving transposition of a local flap with fixation to the sternal area and breast implant inclusion. The other patient who underwent tissue expansion had a breast implant placed and also underwent surgery of the contralateral breast to achieve symmetrization.

Of the remaining four patients, two underwent local flap transposition and breast implantation, with one also undergoing reduction of the contralateral breast. The other two patients were treated only with breast implantation for temporal symmetrization until gland development was complete before the final treatment (Figures 3 and 4).

Three patients with third-degree PS underwent surgical treatment, receiving tissue expanders as an initial approach with the same protocol used in the treatment of patients with second-degree PS. After tissue expansion was complete, each patient underwent a transposition of the major dorsal flap with fixation to the sternal area; two received breast implants and one had a customized implant. Symmetrization of the contralateral breast was performed in two patients (Figure 5).

The symmetrization procedures were required because the aesthetic result was a critical factor in these procedures. All told, seven patients underwent contralateral breast reductions; four of them presented with contralateral mammary hypertrophy. The chosen surgical procedure and the scheduling of the intervention depended on specific conditions for each patient. The final decisions were made with the aim of correcting both the functional and/or aesthetic deformities.

**DISCUSSION**

From the time that Poland described PS in 1841 until 1990, only 400 cases of this syndrome were reported.1 Published studies included the series of seven cases by Mace et al,23 a case report by Samuels et al5 focusing on imaging studies, a study by Merlob et al15 of real-time echo-Doppler duplex scanner evaluation in four cases, the surgical reconstruction of severe cases in four patients by Haller et al,36 and a long-term follow-up of two cases with early correction of the tho-
racic deformity using latissimus dorsi muscle flaps by Anderl and Kerschbaumer. Other reports have included congenital studies of a single patient, like the one by Miller and Miller, and associations of the disease with non-Hodgkin lymphoma, in three cases by Sackey et al.

Although PS is a relatively rare disease according to the literature, we were able to present a large number of definitively diagnosed cases in 28 carefully-evaluated female patients. This number of patients would be equivalent to 7% of the 400 cases reported before 1990 and would also represent a significant proportion of the cases described in other publications since that time.

Beals and Crawford, Sugiura, Padua-Gabriel et al, Alembik and Stoll, Freire-Maia et al, and Taybi
and Lachman\textsuperscript{11} all reported a higher incidence in males than in females. However, Samuel et al\textsuperscript{15} asserted that this condition occurs approximately equally in males and females, and equally on the right and left sides. From the publications of Perez Aznar et al,\textsuperscript{34} Parano et al,\textsuperscript{40} Azzolini et al,\textsuperscript{41} Sackey et al,\textsuperscript{39} Shaham et al,\textsuperscript{42} and Armendares,\textsuperscript{43} we found that most published case reports describe female patients. We chose to review only female patients in our study so that we could standardize the classification system with regard to breast anatomy. Only two male patients were identified and excluded from this study. Future studies are necessary to apply a similar classification system to male patients.
Hurwitz et al.\textsuperscript{22} noted that PS is characterized by different degrees of mammary hypoplasia and alterations to the subcutaneous tissue and adjacent muscles, except the skin and pleural membrane. Patients with such disease have a risk of lung herniation during respiration.\textsuperscript{31} Syndactyly may also be present.\textsuperscript{22} All of these components are not necessarily present, but the full syndrome consists of an assortment of abnormalities.\textsuperscript{14}

In this series, the general characteristics of PS were observed in all 28 female patients. The patients had different degrees of involvement of the chest wall and superior limbs; many had congenital abnormalities associated with PS. The CRPS classification organized these findings into the three degrees of severity described above.

The CRPS classification seems to help in the evaluation needed before beginning any intervention for PS. This classification defines the disease in detail; each degree represents specific deformities defining the main tissues requiring reconstruction, which facilitates the practical and effective study of its pathology in female patients. The classification also allows for a correlation of the clinical manifestations with the imaging tests, making it useful for surgeons in deciding on the best reconstructive procedure. In our clinical experience, it is a clear and easy system that even inexperienced surgeons in plastic surgery departments could use. Without the imaging studies, an accurate evaluation of the disease would not be possible.

Perez Aznar et al.\textsuperscript{34} showed how mild forms of PS are more frequent than severe forms and may go undiagnosed. Hypoplasia of one breast or a horizontal anterior axillary fold may be the sole clinical manifestation of this syndrome.\textsuperscript{44} In our cohort, we observed the severe type (second degree) more frequently than the other types, as it constituted 14 cases. However, most of the 10 patients with mild disease were referred to a specialist for aesthetic reasons.

Martin and Emory\textsuperscript{45} were the first to report macro-mastody associated with PS. A patient with grade III/IV mammary hypertrophy was studied using anamnesis and preoperative physical and imaging examinations. In this case, the surgical treatment confirmed the absence of the PMM.\textsuperscript{45} In our cohort, four of the patients who underwent surgery presented with contralateral mammary hypertrophy and required surgical intervention to achieve symmetrization of the breasts.

Radiographic evaluation of the most significant deformities usually includes a chest radiograph, stereophotogrammetry, and CT and MRI scans, the latter two of which can be used to define the chest wall deformity more accurately once it has been diagnosed.\textsuperscript{46} The radiologic imaging techniques have a fundamental role in the detection, localization, and characterization of thoracic alterations.

Even though the chest radiograph has been the basic imaging study for PS, CT may be required to reveal the total absence of the PMM.\textsuperscript{1} In addition, MRI has made the study of the syndrome much easier because of its high resolution and capacity to generate multiplanar images.\textsuperscript{35} Conventional CT can allow the differentiation of PS from other entities and may facilitate the recognition of various radiologic findings, improve the accuracy of diagnosis, and help optimize treatment.\textsuperscript{32} The images seen on the helical CT scan allow for an easier approach to the disease, even for the nonspecialist members of the reconstruction team. For these and other reasons, this study included all of the available imaging examinations. MRI was indicated for the prospective part of this study; before it became available, radiography and CT scan were the only imaging modalities that could provide such data. Other imaging examinations, including helical CT scan of the thorax and 3-D reconstructions, were performed in severe cases.

The treatment of this condition is strictly surgical.\textsuperscript{22–29,31–33,44,47} The indication for surgery and the time of intervention depend on the severity of the malformations. Fonkalsrud\textsuperscript{48} reported that if surgical repair was performed at an early age, there was a high recurrence rate because of later periods of rapid bone growth. In this study, a cohort of teenagers was studied for surgical treatment; because their development was not complete, temporary interventions (such as the use of tissue expanders) were performed. The earliest age for first stage treatment was 15 years. All severe and highly severe cases were referred to thoracic and orthopedic specialists before any treatment was performed; the opinion of the specialist made the surgical choice easier.

**CONCLUSIONS**

The literature indicates that radiologic diagnosis of thoracic cage alterations is indispensable for planning the treatment and defining a precise surgical approach for PS. Based on the analysis of the imaging studies and clinical evaluation of 28 cases using the CRPS classification, an accurate study was possible for each case, adding valuable information that could be used in deciding upon the surgical approach. The classification also includes a treatment algorithm for each level of disease. The surgical procedure chosen and the time of intervention depended on the details of each case and the final decisions were made with the aim of correcting the functional and/or aesthetic alterations.

During the study, the researchers found that their proposed system of classification was a practical and useful tool that facilitated the choice of a surgical approach. This study only included female patients; further research is required to develop and evaluate a similar classification system for PS in males.

**DISCLOSURES**

The authors have no disclosures with respect to the contents of this article.
Figure 4. A, C, E, Preoperative views of a 20-year-old woman with severe, second-degree Poland syndrome affecting the left side. Note the absence of the pectoralis major muscle, amastia, and nipple–areolar complex asymmetry. B, D, F, Sixty days after placement of the implant, showing good volume, breast symmetry, and filling of the entire hemithorax.
REFERENCES


Figure 5. A, C, Preoperative views of a 13-year-old woman with very severe, third-degree Poland syndrome affecting the right side. Note the severe deformity, breast and nipple–areolar complex asymmetry, total absence of the pectoralis major and minor muscles, sternum rotation, absence of the axillary fold, and osteoarticular deformity in the thoracic cage. B, D, Postoperative views of the same patient in parts A and C at age 28, after tissue expander placement (round, 400 cc), later replaced with a round, high-profile 260-cc cohesive silicone gel–filled implant using a latissimus dorsi muscle flap for coverage. An areola graft was used from the left areola. Contralateral reduction with an inverted-T incision was performed in order to achieve symmetry.