

## I. BACKGROUND/RESEARCH

Non-cancerous breast deformities may occur in men or women and result from a variety of conditions including congenital errors, trauma, disease or aging. Some of the more common deformities that require breast reconstruction surgery include Poland's Syndrome and tuberous breast(s).

Poland's Syndrome is characterized by unilateral aplasia of the pectoralis major muscle and/or minor muscle with ipsilateral symbrachydactyly<sup>1,2</sup> and in some cases, hypoplastic bone and cartilage deformities. Pectus excavatum and pectus carinatum involve either depression or prominence respectively of the sternum and adjacent cartilages. These manifestations may present with athelia, amastia and/or abnormal positioning of the breast and nipple areolar complex.<sup>3</sup> Tuberous breast deformities result from hypoplasia of part or all of the lower quadrant of the breast and can present with skin deficiency and breast constriction.<sup>4,5</sup>

Less common breast deformities include supernumerary breasts and nipples that may present along the mammary ridge that extends from the axilla to the inguinal area bilaterally. Injury or trauma to the chest may result in breast deformity. Aging of the breast or removal of implants may produce severe stretching and loss of upper pole fullness of the breast.<sup>6</sup>

Surgery is the treatment of choice to correct these defects and should be performed when the patient and the surgeon decide it is suitable and the emotional issues relative to growth and development are taken into consideration. Matching the appropriate technique to the appropriate patient is a challenge. Please see other practice guidelines pertaining to reconstruction of the breast after cancer for a complete discussion of the topic.

## II. DIAGNOSTIC CRITERIA

A complete history of the patient's congenital or acquired breast deformity should be obtained. This would include the nature of the defect, as well as other pertinent medical history, especially information regarding prior surgery to the breast or trunk, family history of breast cancer, breast disease, and mammogram results if indicated. The number of pregnancies, live births, and history of nursing should also be determined.

Physical examination of the breast defect should note the size, the configuration (particularly whether there is constriction or a tubular appearance), scarring, and appearance and position of nipple areola complex of the affected breast. The status of the skin and subcutaneous fat should be evaluated. The contralateral breast should be assessed for symmetry and include the degree of ptosis and striae if present. The pectoralis major muscle should be palpated on relaxation and forced contraction. The position of the shoulder, rib cage, and the sternum should also be examined. The scapula and the spine should be observed and the degree of scoliosis, kyphosis and rotation of the chest documented. In case of severe pectoralis excavatum, the cardiac and pulmonary status can be

compromised therefore a cardiology or pulmonary consult may be indicated. In addition, a CT scan and MRI may be necessary to evaluate the muscular status of the chest wall. In rare cases when vascular abnormalities are suspected, a color coded duplex sonography or a contrast enhanced MR angiography may be indicated.<sup>7</sup>

## III. TREATMENT

The surgical technique utilized is dependent on the nature of the defect and the overall health of the patient. The opposite breast must be considered in the initial plan and any underlying deficiencies corrected in order to achieve breast symmetry. When planning for surgery the patient's desired body habitus, occupation, and recreational and sports activities must be taken into considered.

### *Implant/Expander*

An implant is a good choice to fill a defect caused by too little breast tissue. However, there must be sufficient soft tissue to cover the implant. If the skin is constricted, a soft tissue expander may be used to stretch the skin. The expander is placed beneath the skin or chest muscle and over time injected with saline to expand it. Once a sufficient skin envelope is achieved, the expander is removed and replaced by a permanent implant. These surgeries are performed as outpatient procedures.<sup>8</sup>

A solid customized silicone implant can be placed subcutaneously in a mild case of Poland's Syndrome in a male.<sup>9,10,11,12</sup> Usually the prosthesis is based on a presurgical moulage. A drain may be placed into the lateral portion of the pocket and then removed in a few days.

### *Muscle/Musculocutaneous Flaps*

With an anomaly of the pectoral muscle, the use of muscle or musculocutaneous flaps can correct the muscular deficiency. The flap may be either rotational or free flap<sup>13,14</sup> and may be performed either by a standard approach or endoscopically under general anesthesia in a hospital operating room.

The latissimus dorsi muscle flap is a common rotational flap utilized in breast reconstruction and in the case of Poland's Syndrome, it is used to fill the defect created by the lack of the pectoralis major muscle. The latissimus dorsi muscle flap can be harvested endoscopically limiting the skin incision. If there is also a skin deficiency, the skin superficial to the entire muscle can be included in the flap.

When the latissimus dorsi flap is unavailable, the transverse rectus abdominis muscle or TRAM flap can be used. The TRAM flap may be rotated or transferred as a free flap.



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## Correction of Deformed Cartilages

Resection of the deformed cartilages in pectus excavatum or pectus carinatum may be necessary to ultimately produce a chest configuration on which to base the remaining procedures. The sternum is bent forward to an over-corrected position and held in place. In severe cases of pectoralis excavatum, the pectoralis major muscles may be rotated toward the midline. Appropriate consultation with a pediatric or thoracic surgeon may be warranted.<sup>3,15</sup>

## Correction of Tuberos Breast

The surgical correction of the tuberous breast is dependent upon the patient's anatomy and the degree of asymmetry present. The tuberous breast classification listed below is based on a modification of the classification of Von Heimburg, et al.<sup>4,16,17,18,19</sup> However, it should be noted that many deformities are complex and are not categorized into discreet classifications. As such, various treatment plans may be appropriate, depending on the nature of the defect. In some cases, two separate procedures may be necessary to correct the defect. Even in these circumstances it may not be possible to completely correct the defect or to achieve asymmetry.

### Tuberous Breast Classifications

**Type I:** The lower medial quadrant of the breast is deficient but the lower lateral quadrant is ptotic with excess skin.

Treatment: A dermoglandular flap of tissue from the lateral portion of the breast is rotated to fill the medial area. An implant may also be placed subglandularly or submuscularly.

**Type II:** The entire lower quadrant of the breast is deficient of tissue as well as a deficiency of skin in the subareolar area.

Treatment: The upper pole breast tissue is divided to reshape the breast. If breast volume is inadequate, an implant is placed either subglandularly or submuscularly.

**Type III:** The breast base is constricted with an underdeveloped gland. Herniation of the areola is present in greater than 75 percent of cases.

Treatment: The insufficient breast tissue must be redistributed to reshape the breast. An implant or soft tissue expander is placed submuscularly and the inframammary crease is repositioned. The areola is reduced in size and the herniation is reduced, which may be done as a second stage reconstruction.

## Ptosis Correction

If glandular ptosis of the breast exists, placement of an implant subglandularly or submuscularly will lift the breast. If correction cannot be achieved with an implant alone, skin around the areola may be resected.<sup>20,21</sup>

For more significant ptosis where the nipple position is at or below the inframammary crease, the redundant skin is excised. Internal breast flaps may be used to improve symmetry and the areola diameter may be reduced at the same time.<sup>6,22</sup>

## Provider Qualifications

The individual performing this procedure, regardless of the location of the surgical facility should have fully approved hospital privileges for this procedure and be qualified for examination or be certified by a surgical Board recognized by the American Board of Medical Specialties® such as The American Board of Plastic Surgery, Inc.®

## IV. DISCLAIMER

Practice parameters are strategies for patient management, developed to assist physicians in clinical decision making. This practice parameter, based on a thorough evaluation of the scientific literature and relevant clinical experience, describes a range of generally acceptable approaches to diagnose, manage, or prevent specific disease or conditions. This practice parameter attempts to define principles of practice that should generally meet the needs of most patients in most circumstances.

However, this practice parameter should not be construed as a rule, nor should it be deemed inclusive of all proper methods of care or exclusive of other methods of care reasonably directed at obtaining the appropriate results. It is anticipated that it will be necessary to approach some patients' needs in different ways. The ultimate judgment regarding the care of a particular patient must be made by the physician in light of all the circumstances presented by the patient, the diagnostic and treatment options available and available resources.

This practice parameter is not intended to define or serve as a standard of medical care. Standards of medical care are determined on the basis of all the facts or circumstances involved in an individual case and are subject to change as scientific knowledge and technology advance and as practice patterns evolve. This practice parameter reflects the state of knowledge current at the time of publication. Given the inevitable changes in scientific information and technology, periodic review, updating and revision will be done.

## V. CODING

### ICD-9

A. Deformity of the breast (acquired)	611.8
B. Absence of breast(s), congenital	757.6
C. Malignant neoplasm female breast	174
Nipple and areola	174.0
Central portion	174.1
Upper-inner quadrant	174.2
Lower-inner quadrant	174.3
Upper-outer quadrant	174.4
Lower-outer quadrant	174.5
Axillary tail	174.6
Other specified sites of the breast	174.8
Breast, female, unspecified	174.9
D. Family history malignant neoplasm, breast	V16.3
E. Personal history malignant neoplasm, breast	V10.3
F. Specified anomalies of breast	757.6
Absent	
Accessory, breast or nipple	
Supernumerary	
Hypoplasia of breast	

<b>Procedure</b>	<b>CPT</b>
A. Mammoplasty, augmentation; without prosthetic implant with prosthetic implant	19324 19325
B. Immediate insertion of breast prosthesis following mastectomy or in reconstruction	19340
C. Delayed insertion of breast prosthesis following mastopexy, mastectomy or in reconstruction	19342
D. Nipple/areolar reconstruction	19350
E. Breast reconstruction, immediate or delayed, with tissue expander, including subsequent expansion	19357
F. Breast reconstruction with latissimus dorsi flap, with or without prosthetic implant	19361
G. Breast reconstruction with free flap	19364
H. Breast reconstruction with other technique	19366
I. Breast reconstruction with transverse rectus abdominis myocutaneous flap (TRAM), single pedicle, including closure of donor site	19367
double, pedicle, including closure of donor site	19369
J. Open periprosthetic capsulotomy, breast	19370
K. Periprosthetic capsulectomy, breast	19371
L. Revision of reconstructed breast	19380
M. Preparation of moulage for custom breast implant	19396
N. Unlisted procedure, breast	19499

## VI. REFERENCES

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