Description

Poland Syndrome
Includes the features of ipsilateral breast and nipple hypoplasia and/or aplasia, deficiencies of subcutaneous fat and axillary hair, absence of the sternum head of the pectoralis major, hypoplasia of the rig cage, and hypoplasia of the upper extremity.

Patient with disorder can present with ipsilateral involvement of the chest muscles, skin and subcutaneous tissues, bones, and upper extremity. The absence of the sternum head of the pectoralis major muscle is considered the minimal expression of the syndrome. Involvement of adjacent muscles, including the pectoralis minor, serratus, latissimus dorsi, and the external oblique, also has been described.

The skin of the area is hypoplastic with a thinned subcutaneous layer and the axillary hair may be absent. The nipple is often smaller and higher in both males and females and the breast is generally hypoplastic in females. Skeletal deformities may involve absence of portions of the ribs or costal cartilages anteriorly. In severe cases, anterior lung herniation may be present. The scapula may be smaller with winging (Sprengel) deformity. The upper extremity also may be hypoplastic, the upper arm, forearm, and fingers may be shortened (brachysymphalangism).

Reconstructive Surgery
Refers to surgical procedures to rebuild the contour of the breast, along with the nipple and areola if desired. Reconstructive surgery is performed on abnormal structures of the body, caused by congenital defects, developmental abnormalities, trauma, infection, tumors, or disease. It is generally performed to improve function, but may also be done to approximate a normal appearance.

Cosmetic Surgery
Is performed to reshape normal structures of the body in order to improve the patient’s appearance and self-esteem.

Decision Criteria

Administrative
1. Referral from affected member’s PCP along with supporting medical documentation justifying need for proposed surgical procedure. Appropriate medical documentation should include:
   a. Member’s name & THC ID#
   b. Proposed procedure; ICD, CPT, or other appropriate codes; surgical date, name of facility, surgeon, etc.
   c. Name and contact numbers for treating provider and facility
d. Progress notes containing appropriate and relevant documentation

e. Photographs (recommended but not required)

f. Diagnostic studies (CT scan of chest, EKG if heart murmur or known heart disorder; chest x-ray, and appropriate lab studies to rule out malignancy as cause) documenting deformity and results

g. Cardiopulmonary studies documenting respiratory/cardiac impairment

2. Services must be performed by a THC affiliated or contracted provider (physician, hospital, or other health care provider), unless approved in advanced by Plan Medical Director for OON services.

3. Procedure must be prior authorized by Plan or Medical Director

4. Member must have current eligibility on date of service

5. Procedure must be determine to be medically necessary and not considered cosmetic in nature

Clinical

1. Clinical documentation of Poland Syndrome as evidenced by CAT scan of chest documenting deformities associated with the disorder. CT of chest documents pectus index is >3.25

2. Congenital absence or hypoplasia of pectoralis major and minor muscles; breast hypoplasia, and congenital partial absence of the upper costal cartilage

3. Well documented evidence of complications arising from the sternal deformity including but not limited to:
   a. Cardiopulmonary impairment documented by respiratory and/ocardiac function tests
   b. Frequent lower respiratory tract infections
   c. Asthma
   d. Exercise limitation
   e. Atypical chest pain
   f. EKG or echocardiogram if a heart murmur or known cardiac disorder is present to define the relationship of the cardiac problem to the sternal deformity
   g. Children should be older than 4 years with males preferably at least 13 years of age.
   h. Reconstruction should not be performed too early in children to prevent asymmetry as a result of continued growth of the breasts, necessitating a secondary revision procedure once breast development is completed
   i. Person must be accept to maturely accept the responsibility of a foreign body and the ability to deal with potential complications (further surgery for tissue expansion)

Bibliography


Medical Decision Criteria-Surgical Indications for Poland Syndrome


4. emedicine from WebMD; “Breast, Poland Syndrome”. Author: Bradon J. Wilhelmi, MD; Leonard Weiner, MD; Professional, Chief of Plastic Surgery, University of Louisville. Stanford University Medical Center. Pages 1-9. http://www.emedicine.com/plastic/topic132.htm; accessed 07/17/07