Polland Syndrome

OVERVIEW
Your child has been diagnosed with Polland Syndrome. This condition was named after Sir Alfred Poland, who described it in 1841. This term describes a congenital condition in which the child is born without one of the pectoralis muscles. There can be underdevelopment of skin, breast, adjacent muscles, rib cage and arm.

BACKGROUND
This condition is uncommon, but not rare. It is under reported so the incidence is difficult to determine, but one review estimated the incidence at 1 in 30,000. The right side is affected twice as often as the left. Females seek treatment more than males, but there is no known gender predilection. Most cases arise sporadically. However, there are several reports of family members and twins, suggesting some degree of genetic transmission. It has been associated with other syndromes including Möbius syndrome (congenital bilateral facial paralysis with inability to abduct the eyes) and Klippel-Feil syndrome. Hematopoietic malignancies, including leukemia and non-Hodgkin lymphoma, have been described.

Several theories have been advanced to explain the etiology of Poland syndrome, most evidence indicates that it results from a vascular event during the critical sixth week of gestation with hypoplasia of the subclavian artery causing the musculoskeletal malformations.

DIAGNOSIS
Because the functional disability in Poland syndrome is mild, patients usually present as older children for evaluation and discussion of the chest and shoulder shape. This is typically managed by plastic surgeons, who can provide more details. During the examination, the stage of breast development is noted and the status of the latissimus dorsi muscle is checked. Extraocular muscle motion must be confirmed in excluding Möbius syndrome. Examination of lymph nodes and complete blood cell count should assist with evaluation for associated leukemia and non-Hodgkin lymphoma. A chest radiograph may be indicated to determine rib abnormalities or the presence of diaphragmatic hernia. When reconstruction is considered with a plastic surgeon for cosmetic reasons, a CT scan is sometimes used to clarify the shape of the chest wall and to determine the presence of the latissimus muscle.
TREATMENT
Patient is usually for the chest deformity and breast asymmetry. Females often benefit from staged breast augmentation with a subcutaneous tissue expander filled with fluid that can expanded at weekly intervals until the affected breast matches the contralateral side and expanded further as the unaffected breast grows. After age 18 and completion of breast development, the tissue expander can be removed and reconstruction done with the latissimus muscle transposed over a permanent implant. Occasionally, the nipple will need additional reconstruction. Sometimes the opposite breast, may need reduction or augmentation for optimal symmetry. If the latissimus muscle is absent, other muscles can be used. For males the latissimus muscle can be transposed after age 13, to fill the void of the absent pectoralis major muscle. Reconstruction for males with a prosthesis has been described but with disappointing results.

PROGNOSIS AND OUTCOME
Prognosis is good. Treatment is typically directed at improving the appearance of the chest. With appropriate timing, procedure selection, and preoperative evaluation, excellent results and high patient satisfaction can be achieved in the treatment of chest and breast deformities resulting from Poland syndrome.

MORE INFORMATION
Further information can be obtained on the internet. Your local public library can help you explore these sources if you are interested. Two good sites for expert and peer reviewed information are the American Academy of Orthopedic Surgeons at www.aaos.org and www.emedicine.com.

FEEDBACK
If you have questions or comments, please contact the office or submit them to the web site at www.pedortho.com.