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# Poland's Syndrome

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Poland's Syndrome is a congenital deformity consisting of ipsilateral syndactyly and pectoral girdle muscle deficiency. An illustrative case of this anomaly is presented along with a literature review.

**Illustrative Case**

On the newborn physical examination, a 5 lb. 13oz. full term infant female exhibited hypoplasia of the left hand with syndactyly of the index and middle fingers to the level of the PIP joints (fig. 1), absence of the sternocostal head of the pctoralis major (fig. 2), and overlapping of the fifth toes bilaterally (fig. 3).

There was no family history of syndactyly, absence of the pectoralis major, or other congenital anamolies. The mother had a long history of infirtility for which she had been prescribed clomiphene citrate without results. This pregnancy occured three years after discontinuing clomiphene with no further medical or surgical intervention. She did not realize she was pregnant until the end of the first trimester, and during that time consumed aspirin and smoked one pack per day. She denied use of alcohol or other drugs. The only prenatal complication noted was a mild intrauterine growth retardation which was attributed to the mother's use of tobacco.

The infant was referred to a plastic surgeon who deferred plastic surgical correction of her syndactyly untill she reached one year of age, and to an orthopedic surgeon who followed her overlapping toes and will surgically release the extensor tendons if resolution does not occur in the first year. Reconstruction of the breast and cest wall will be considered at approximately 15 years.

**Discussion**

In 1841, Alfred Poland performed an autopsy on a 27-year-old convict which revealed absence of the pectoralis minor with hypoplasia of the sternocostal portion of the pectoralis major, serratus anterior, and abdominal oblique externa. The hypoplastic ipsilateral hand displayed syndactyly of the four fingers to the level of the PIP joints with agenesis of three of the middle phalanges and hypoplasia of the fourth. The thoracic vessels supplying the intercostal spaces were described as very small.

Since Poland's original description, several cases have been published of the association of ipsilateral thoracic and hand anamolies which has come to be known as Poland's syndrome. The severity and extent of the chest and hand involvement varies. The thoracic aberration may include hypoplasia or absence of the pectoralis major, pectoralis minor, deltoid, serratus anterior, external oblique, and latissimus dorsi, abnoralities of the ribs, clavicle and sternum, lung hernation, Sprengel's deformity, scoliosis, hemivertebrae, dextrocardia, and absence or underdevelopment of the breast. The upper extremity deformity may be characterized by syndactyly, hypoplasia or absence of metacarpals or phalanges, absence of extensors or flexor tendons, carpal coalition or hypoplasia, shortening of the humerus, radius, and/or ulna, radioulnar synostosis, and nail agenesis. Associated anomalies include foot deformaties, Mobius syndrome, Bonnevie-Ullrich syndrome, thrombocytopenia, leukemia, lymphona and spherocytosis.

Consequently, evaluation of the patient with Poland's syndrome should include, in addition to a

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thorough physical, a radiograph of the chest and a complete blood count.

The incidence of Poland's syndrome is 1:32,000 live births and is found in 10% of patients with syndactyly. Syndactyly alone is found in 1:2,500 live births. The syndrome is tmore common in males than in females, and is right side in 75% of the cases. (However, in our case, a female was affected on the left side.)

The etiology of the syndrome is unknown, but it has been theorized that a vascular malformation delivers insufficient blood to the developing shoulder girdle and limb at the critical period in the limb bud development, approximately seven weeks gestation. Arterial anamolies and stenoses have been seen on arteriographs. A case of Poland's syndrome was reported in which the mother attempted to induce an abortion at seven weeksgestation by ingesting a large number of ergonovine maleate tablets, which theoretically may have produced vascular spasm in the developing limb bud. In David's report of 10 cases of Poland's anomoly, five mothers admitted or were strongly suspected of attempting abortion early in pregnancy. Theoretically, any resultamt plaacental separation may have produced fetal hypoxia, and short-term hypoxia has been shown to cause skeletal defects in animals. In the other five cases, the infants were adopted, thus a maternal history was unavailable. Boaz et al. reported a case of Poland's syndrome in which the prenatal history was marked by severe maternal bleeding at approximately six weeks.

Most cases reported appear not to be genetically determined. However, the case of two siblings, one with the "full syndrome" and the other with only syndactyly and absence of the pectoralis major, has been reported.

**Management**

The patient with PS who exhibits syndactyly should be referred to a plastic surgeon. Surgical bony separation initiated within the first year of life is felt by many to achieve a better functional result. Consequently, separation is usually performed at one year of age when the child presents less of an anesthetic danger, yet retains an abundance of fat around the dorsum of the hand. The fat serves as a natural tissue expander and its reemoval allows the surgical wound edges to be more closely approximated. However, complete closure usually necessitates a full thickness skin graft from an area such as the groin.

Complications include graft failure, hair growth in the graft, flexion contractures, and recurrent syndactyly (poor bandaging resulting in the juxtaposition of raw interdigital surfaces with adhesion formation and resultant iatrogenic syndactyly). Fortunately, 95% of patients suffer no complications. Redeepening of web spaces is often required after the pubertal growth spurt.

The thoracis deformity usually presents a cosmetic imperfection rather than a fuctional disability. However, severe forms may result in diminished strength in the affected girdle. Anderl and Kerschbaumer recommend early plastic surgery when signs of thoracic cage deformation become evident. Although little deformity may be present at birth, they feel that a continuous process of deformation occurs over several years as the costal cartilages are elevated and the sternum is twisted by the strong force of the pectoralis muscle on the healthy side.

the thoracic deformity is corrected by transposition of a latissismus dorsi muscle flap. The muscle flap is raised dorsally and transposed anteriorly in order to simulate the absent anxillary fold. Postpubertal females may benefit from a submuscular breast augmentation (and possible contralateral breast reduction). Reconstruction by the use of rib grafts or custom-made chest walls prostheses is less favorable since local tissue erosion, migration and adverse cosmesis often result.

In summary, Poland's syndrome is acongenital aberration of the shoulder-limb girdle and hand which exhibits a varying degree of severity, and in most cases poses no functional impairment, but only adverse cosmesis for which plastic surgery is most helpful.