The Association for Children with Hand or Arm Deficiency

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Poland's Syndrome Information Sheet

(Also known as Poland Anomaly, Poland complex and Poland's Syndactyly)

A brief description

Unilateral absence of the pectoralis muscle and ipsolateral anomalies of the hand (although in rare cases the hand may be unaffected).

The most common physical signs of this syndrome are a missing chest muscle, combined with an imperfectly formed hand. The hand may be 'small' with webbing of the fingers and sometimes the thumb. Less frequently the radius bone may be affected.

The majority of cases are sporadic, isolated events. Apart from the physical difference the child is otherwise 'normal'. The defect is rare effecting possibly as few as 1 in 25000 births, that is approximately 20 babies each year in the UK.

Poland's syndrome can affect either side of the body but seems to occur more often on the right side. It would appear to be more common in boys than girls.

Extract taken from – 'The Malformed Infant and Child', RM Goodman &RJ Gorlin. 1983

Clinical Features: The Poland anomaly or developmental field defect is clinically variable. It consists of unilateral absence of the sternal and costal portions of the pectoralis major muscle, and symbrachydactyly of the hand on the ipsilateral side. In its full expression there are hypoplasia of the skin and subcutaneous tissues of the anterior chest, absence or hypoplasia and upward displacement of the nipple and breast, pectoral and axillary hypotrichosis, absence of the sternocostal portion of the pectoralis major, absence of the pectoralis minor (one-third as often), absence of portions of costal cartilages two, three, and four or three, four, and five; absence of the middle phalanges of all the fingers or fusion of the middle and distal phalanges with webbing between the proximal phalanges. There is no bony synostosis. The hand on the ipsilateral side is usually smaller than that on the unaffected side. The thumb is often normal. Occasionally other muscles about the shoulder girdle have been deficient, and Sprengel deformity has been noted in some cases.

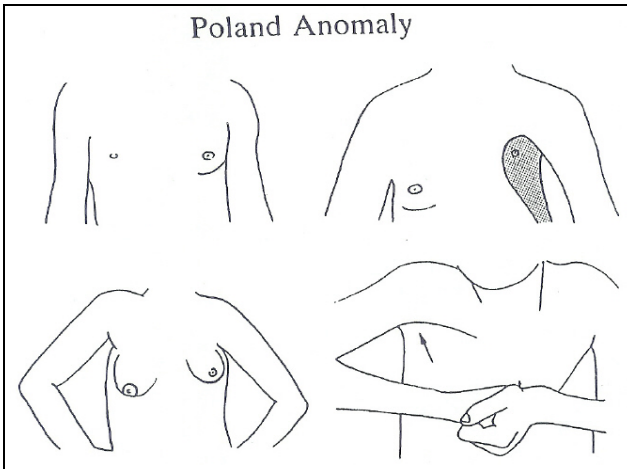
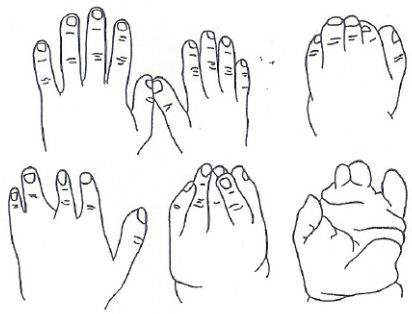
Specific DiagnosIs The diagnosis is a clinical one. The condition has occurred without defects of the hand, without costal defects, and without recognizable defects of other muscles of the shoulder girdle. However, the virtually constant features are anomalies of the nipple, of the subcutaneous tissues. and of the pectoralis major and minor. Webbing of the axilla is not uncommon. Moebius syndrome exhibiting paralysis of the sixth and seventh cranial nerves may be associated with, among other defects, the hand and chest wall features of Poland anomaly in approximately 15% of cases.

Differential Diagnosis The disorder is so characteristic that discussion of differential diagnosis is not relevant.

Prenatal Diagnosis When the limb and chest deformities are severe, ultrasound studies and possibly fetoscopy could be used to diagnose this condition.

Basic Defect, Genetics and Other Considerations The disorder is not hereditary. The primary defect appears to be a local one in mesoderm in the quadrant from the pectoral area to the distal upper limb. It has been estimated that approximately 10% of patients with syndactyly of the hand have the Poland anomaly. Approximately 75% of patients with Poland anomaly are male and in about 70% the right side is involved. Most cases have been sporadic. Various estimates concerning its frequency have ranged from 1 in 7000 to I in 100,000. However, this may not reflect the true frequency if the hand anomalies are mild. There have been several familial examples, most involving parent and child. But the recurrence rate is considerably less than 1 %.

Prognosis and Treatment The chest abnormalities are not treated. Correction of syndactyly may be carried out if function may be improved.

	
Unilateral absence or hypoplasia of the pectoralis minor and sternal portion of the pectoralis major (arrow), absence of hypoplasia of nipple and areola	Ipsilateral hand deformities with small hand, brachydactyly, syndactyly and terminal symphalangism

Comments from our medical advisor Mr Simon Kay, FRCS of St James's University Hospital

Poland's Anomaly is a fascinating condition. First described by a medical student called Poland at Guy's Hospital in the latter part of the last century, it is generally used to describe the combination of hand abnormalities with abnormalities of the shoulder girdle and chest wall on the same side. The condition is not hereditary and our present understanding is that it probably occurs from a very temporary alteration in the blood flow through the limb of the very small foetus. Interestingly it is only ever seen on one side or the other unlike most other congenital abnormalities.

The abnormality in the hand may vary from just a mild change in the skin crease patterns of the hand to complete loss of some digits and typically stiffness or webbing of the remaining digits. The whole arm is usually shorter and slightly smaller and the most common abnormality on the shoulder girdle and chest wall is for the muscle in the region of the front of the chest that passes from beneath the breast to the arm and makes the anterior fold of the armpit, (the pectoralis major muscle) to be small or absent. This usually is combined with a small breast with a small nipple complex.
My regiment for this condition has evolved over the years and has changed with the advent of new technology.

In the hand the variability of the conditions found means that no one treatment is appropriate to all but it is most unusual with modern techniques not to be able to achieve useful hand function.

In the shoulder girdle, I usually like to transpose a muscle from the back of the chest to the front because the lack of a fold at the front of the armpit is quite unsightly. Then during adolescence I gradually stretch the skin of the breast with what is known as a tissue expander (balloon) beneath the breast skin. When breast enlargement ceases one can replace the expander with either an implant or nowadays there are techniques for replacing it with living tissue which gives a more natural feel and shape to the breast. It may on occasions be necessary to reduce or lift the opposite breast at some stage in order to match shape and size more accurately.

Further Information

For further information contact the Poland Support Group.

The POLAND SYNDROME Support Group is here to give
Support, Advice and Information to anyone with Poland Syndrome, their Families and Friends.
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