

# *Poland Syndrome Thorax*

## **Treatment of Poland syndrome thorax deformity with the lipomodeling technique: About ten cases].**

[Article in French]

[La Marca S](#), [Delay E](#), [Toussoun G](#), [Ho Quoc C](#), [Sinna R](#).

### **Source**

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### **Abstract**

The correct management, with the classic techniques, of the thoracic deformity in Poland's syndrome is difficult, with often insatisfactory results. The current surgical treatment involves the use of prothetic material and/or different flaps with their own complications and scares. The experience of our team with fat grafting (we named lipomodeling) in breast reconstruction helped us to propose the correction of the thoracic and mammary deformity by repeated fat transfer sessions. Fat grafting is commonly used in our team since 1998 in various indication of breast surgery. We reviewed retrospectively our ten first cases of thoracic deformity in Poland's syndrome treated with only fat grafting. Patients had repeated procedures until obtaining a satisfactory result. The fat was harvested from the thigh, buttocks, and abdomen. There were young patients with a mean age of 16years old (from 12 to 24). The mean follow-up was 51months. An average of 2.9 procedures (1 to 5) with 255cm(3) of fat injection at each procedure was needed to obtain symetry. Hundred percent of the patients were satisfied. No complication was noted. As reported, the reconstruction of the thoracic deformity and the mammary shape can be obtained by fat grafting. The absence of a flap donor site sequelae and the absence an implant allow this technique to be simple, reproducible, and without great complication. These criteria match well the surgical management of this deformity, which is mainly aesthetic. Moreover, the secondary benefit of liposuction of disgracious steatomery helps the acceptance of the procedure. Therefore in our hands, fat grafting to the breast (lipomodeling) is now our first choice treatment in thoracic Poland syndrome deformity. Given the rarity of this syndrome, we recommend a treatment by an operator who makes the learning curve of lipomodeling, and who often deals with Poland syndrome.

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## **Breast and chest wall reconstruction with the transverse musculocutaneous gracilis flap inPoland syndrome.**

[Huemer GM](#), [Puelzl P](#), [Schoeller T](#).

### **Source**

Department of Plastic and Reconstructive Surgery, Marienhospital Stuttgart, Stuttgart, Germany.

### **Abstract**

Poland syndrome is a complex chest wall deformity with unilateral hypoplasia of the breast and pectoralis muscle, with a missing anterior axillary fold in its most common form. The authors report their combined experience and technique with the transverse myocutaneous gracilis flap

to reconstruct the chest wall and breast either alone or simultaneously. Between June of 2004 and July of 2010, 11 patients (two male patients) were operated on and 14 flaps were transplanted. The authors found that the transverse myocutaneous gracilis flap proved to be a very valuable microsurgical alternative for reconstructing the chest wall and female breast in Poland syndrome with autologous tissue. The flap provides the surgeon maximal freedom of flap inseting for optimal symmetry together with a very inconspicuous donor site regardless of unilateral or bilateral harvesting.

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## **Chest wall repair in Poland syndrome: complex single-stage surgery including Vertical Expandable Prosthetic Titanium Rib stabilization--a case report.**

Lieber J, Kirschner HJ, Fuchs J.

### **Source**

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### **Abstract**

Various surgical techniques have been described for repair of chest wall defects in Poland syndrome. We describe the case of a 16-year-old boy who underwent autologous rib transposition after sternal osteotomy. Chest wall stabilization was achieved using a combination of K-wires and Vertical Expandable Prosthetic Titanium Rib (Synthes GmbH, Freiburg, Germany). Reconstruction of the soft tissue defect was accomplished by combined latissimus dorsi muscle flap and Permacol patch (Covidien Deutschland GmbH, Neustadt, Germany). This approach might be considered an effective 1-stage treatment option of this condition in postpubescent boys.

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## **Muscle abnormalities of the chest in Poland's syndrome: variations and proposal for a classification.**

Stylianios K, Constantinos P, Alexandros T, Aliki F, Nikolaos A, Demetriou M, Petros P.

### **Source**

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### **Abstract**

#### **PURPOSES:**

Poland's syndrome (PS) is a rare congenital malformation, which combines anomalies of the chest and the homolateral upper limb. The purposes of the paper are to study the chest musculoskeletal malformations of the syndrome and propose a classification for the thoracic anomalies through our experience and taking into account the literature.

#### **METHODS:**

We reviewed ten patients diagnosed with PS. All of them suffered from anomalies of the thorax and the ipsilateral upper extremity. We proceeded to scan the thorax with 3-D CT for better imaging of the structures and examination of the variations.

#### **RESULTS:**

All patients were young to middle aged: seven women and three men. Both sides of the body were affected to the same extent. Six patients presented with total absence of the pectoralis

muscles and variable anomalies of the thoracic skeletal structures. Hypoplasia of the minor pectoralis and retraction of the ribs were observed in two patients, while the two other patients presented with major and minor pectoralis absence as well, except for the clavicular head.

#### **CONCLUSIONS:**

There are many variations of PS with regard to the chest that can be best detected with 3-D CT imaging, which should be used whenever it is available. We propose a simple classification of the musculoskeletal anomalies of the chest in Poland's syndrome that would be a useful tool for the clinicians and especially plastic surgeons to make an easy diagnosis. In combination with 3-D CT imaging, surgeons will be able to achieve the best treatment for each patient.

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## **Novel titanium constructs for chest wall reconstruction in children.**

Stephenson JT, Song K, Avansino JR, Meshner A, Waldhausen JH.

#### **Source**

Division of Pediatric Surgery, Seattle Children's Hospital University of Washington School of Medicine, PO Box 5371/ WW7753 Seattle, Washington 98105, USA.

#### **Abstract**

##### **PURPOSE:**

We have previously reported the use of the vertical expandable prosthetic titanium rib (VEPTR) for treatment of thoracic dystrophy. This report describes our experience with this device and other novel titanium constructs for chest wall reconstruction.

##### **METHODS:**

This is a retrospective chart review of all children and adolescents undergoing chest wall reconstruction with titanium constructs between December 2005 and May 2010.

##### **RESULTS:**

Six patients have undergone chest wall reconstruction with VEPTR or other titanium constructs. Four had chest wall resection for primary malignancy, 1 had metastatic chest wall tumor resection, and 1 had congenital chest wall deformity. There were no immediate complications, and all patients have exhibited excellent respiratory function with no scoliosis.

##### **CONCLUSIONS:**

Chest wall reconstruction after tumor resection or for primary chest wall deformities can be effectively accomplished with VEPTR and other customized titanium constructs. Goals should be durable protection of intrathoracic organs and preservation of thoracic volume and function throughout growth. Careful preoperative evaluation and patient-specific planning are important aspects of successful reconstruction.

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## **Poland syndrome: evaluation and treatment of the chest wall in 63 patients.**

Seyfer AE, Fox JP, Hamilton CG.

#### **Source**

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#### **Abstract**

**BACKGROUND:**

Poland syndrome is a sporadic, congenital unilateral absence of the sternocostal head of the pectoralis major muscle that can occur with other ipsilateral chest wall and limb derangements. The chest wall deficiency is primarily cosmetic, its incidence is unknown, male patients may be affected more than female patients, the right side is affected more than the left, and associated comorbidities may exist. Chest wall repair depends on anatomical type and gender.

**METHODS:**

Sixty-three patients with Poland syndrome were divided into two treatment groups by chest wall anatomy and gender. Surgical repair was based on this division. Seventy-six operations were performed by the senior author (A.E.S.) during a 30-year period, and long-term outcomes are presented. Corrective methods included use of custom-made chest wall prostheses, mammary prostheses, latissimus dorsi muscle transfers, transverse rectus abdominis musculocutaneous flaps, sternal/rib reconstruction, or a combination of methods. Follow-up ranged from 1 to 21 years.

**RESULTS:**

Two anatomical forms of the disorder are described, each with unique surgical requirements. The simple deformity was effectively repaired with a latissimus dorsi muscle transfer plus, in female patients, a sublatissimus mammary prosthesis. Repair of the complex deformity, in addition to the latissimus transfer, selectively included musculoskeletal chest wall realignment. Custom-made chest wall prostheses carried a higher risk of complications.

**CONCLUSIONS:**

Poland syndrome of the chest wall exists in two forms: the more common simple variety and a complex form (as originally described by Poland). Repair of the chest wall can be effectively tailored to these anatomical types, gender, and patient preference.

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## **Lipomodeling of Poland's syndrome: a new treatment of the thoracic deformity.**

[Delay E](#), [Sinna R](#), [Chekaroua K](#), [Delaporte T](#), [Garson S](#), [Toussoun G](#).

**Source**

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**Abstract****BACKGROUND:**

The severe forms of Poland's syndrome, with thoracic deformity, were until now very difficult to treat, with treatment involving complex surgery and implant insertion. Results were, in general, inadequate and the appearance unnatural. Our experience with fat transfer for breast reconstruction led us to propose reconstruction of the breast and thorax by serial fat transfer.

**METHODS:**

Our patient had a very severe form of Poland's syndrome with agenesis of the pectoralis major and latissimus dorsi muscles and lack of fusion of the fourth costal arch. She was treated by fat transfer, or lipomodeling. Lipomodeling was developed in our team in 1998 to augment breast volume after autologous latissimus dorsi flap reconstruction. Because this technique and use of an implant were not possible, we attempted reconstruction by repeated lipomodeling. The patient underwent five sessions at intervals of a few months, the first in August 2001.

**RESULTS:**

With 6 years of follow-up, the aesthetic, functional, and psychological results exceeded our expectations. In five sessions we were able to reconstruct a breast of natural shape, sensitivity, and consistency, and which was totally accepted by the patient. Mammography, echography, and MRI 1 year later showed a normal breast of fatty type.

**CONCLUSION:**

Lipomodeling in Poland's syndrome is technically feasible. This original description of treatment of the severest form of Poland's syndrome, with impressive results and at the cost of limited constraints and scar sequelae, opens new perspectives and suggests extensive potential applications of lipomodeling in all disciplines related to the breast.

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**Custom made chest-wall implant and Poland's syndrome: Between art and science.**

[Article in French]

[Perignon D](#), [Marton A](#), [Qassemyar Q](#), [Carton S](#), [Benhaim T](#), [Morez B](#), [Robbe M](#), [Sinna R](#).

**Source**

Department of plastic, reconstructive and aesthetic surgery, Amiens university hospital, Picardie, France.

**Abstract**

Poland's syndrome is a well-described congenital thoracic deformity with mostly only an aesthetic damage. We report the case of a chest-wall correction of a young male patient by a custom made silicone implant, detailing the original process of manufacturing, and justifying the choice not to use a conventional technique of reconstruction. This alternative enhances the surgeon's therapeutic arsenal, so that he can provide a customized answer, adapted to the requirements and the morphological characteristics of each patient.

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**Poland syndrome.**

[Urschel HC Jr](#).

**Source**

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**Abstract**

Poland syndrome is characterized by hypoplasia or absence of the breast or nipple, hypoplasia of subcutaneous tissue, absence of the costosternal portion of the pectoralis major muscle, absence of the pectoralis minor muscle, and absence of costal cartilages or ribs 2, 3, and 4 or 3, 4, and 5. The chest wall defect is often associated with a lung hernia. Clinical manifestations are extremely variable and rarely are all the features recognized in 1 individual. Fortunately it is invariably unilateral, allowing for an easier reconstruction. Single-stage reconstruction of the chest wall combined with simultaneous augmentation mammoplasty and transfer of an island pedicle myocutaneous flap of latissimus dorsi muscle are major improvements over previous multiple-stage procedures that provide less satisfactory cosmetic results in management of patients with Poland syndrome.

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**Anatomical, histologic, and genetic characteristics of congenital chest wall deformities.**

[Fokin AA](#), [Steuerwald NM](#), [Ahrens WA](#), [Allen KE](#).

**Source**

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### **Abstract**

There is a large and diverse group of congenital abnormalities of the thorax that manifest as deformities and/or defects of the anterior chest wall and, depending on the severity and concomitant anomalies, may have cardiopulmonary implications. Pectus excavatum, the most common anterior chest deformity, is characterized by sternal depression with corresponding leftward displacement and rotation of the heart. Pectus carinatum, the second most common, exhibits a variety of chest wall protrusions and very diverse clinical manifestations. The cause of these conditions is thought to be abnormal elongation of the costal cartilages. Collagen, as a major structural component of rib cartilage, is implicated by genetic and histologic analysis. Poland syndrome is a unique unilateral chest/hand deficiency that may include rib defects, pectoral muscle deficit, and syndactyly. Cleft sternum is a rare congenital defect resulting from nonfusion of the sternal halves, which leaves the heart unprotected and requires early surgical intervention.

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## **Simultaneous costal cartilage-sparing modified Ravitch procedure and latissimus dorsi transfer for chest wall deformity repair in Poland's syndrome.**

[Dingeldein MW](#), [Lu CY](#), [Kim AW](#), [Ostric S](#), [Liptay MJ](#), [Holterman MJ](#).

### **Source**

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### **Abstract**

Poland's syndrome is a constellation of rare congenital anomalies that include hypoplasia of breast and underlying subcutaneous tissue, absence of the costosternal portion of the pectoralis major muscle, deformity or absence of ribs, absence of axillary hair, and syndactyly. Various surgical techniques have been described to repair such chest wall defects. We report a case of simultaneous Fonkalsrud procedure (costal cartilage-sparing version of the modified Ravitch procedure) and latissimus dorsi transfer in a 15-year-old boy with Poland's syndrome. The Fonkalsrud procedure has been used in the repair of pectus excavatum and pectus carinatum, and latissimus dorsi muscle transfer has been used in the repair of Poland's syndrome. In this report, we describe their combined use in an adolescent with severe pectus excavatum associated with Poland's syndrome. This combination of established operations resulted in a successful outcome.

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## **Customised silicone prostheses for the reconstruction of chest wall defects: technique of manufacture and final outcome.**

[Saour S](#), [Shaaban H](#), [McPhail J](#), [McArthur P](#).

### **Source**

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### **Abstract**

Chest wall reconstruction with a customised silicone prosthesis in 13 patients (five patients with pectus excavatum, six Poland's syndrome and two with post-surgical chest wall deformity) is presented. An alginate impression or CT scan with three-dimensional reconstruction was used

to produce the final mould from which the silicone prosthesis was fabricated. The surface of the silicone implant was roughened to reduce capsular contracture and holes were incorporated to allow for tissue integration. Twelve patients had aesthetically acceptable results after a mean of 5 years follow up. This series indicates that accurate assessment of the defect, modifications on manufacture and proper placement of the implant result in a more satisfactory final outcome.

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## **Poland's syndrome and hand's malformations: about a clinic series of 37 patients.**

[Article in French]

Foucras L, Grolleau JL, Chavoïn JP.

### **Source**

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### **Abstract**

#### **INTRODUCTION:**

Poland's syndrome is a rare malformation which associates thoracic anomalies and anomalies of homolateral upper end. We wish to know the frequency of hand's malformations in this syndrome in our clinical experience.

#### **MATERIAL AND METHODS:**

We have revised 37 patients who were seen initially for a thoracomammary anomaly. This clinical series from plastic surgery service of Toulouse has been revised to know the importance of hand's malformations.

#### **RESULTS:**

Hand's malformations in Poland's syndrome are rare in your study, they touch only 12% patients. We find only 4 malformations in 33 patients, four were lost. They were only females, we find three brachymesophalanges and a major form.

#### **DISCUSSION:**

Hand's malformations in Poland's syndrome are less frequent than classically. There is no parallelism between gravity of thoracic malformation and that one of upper end. In this series, we find only one case with syndactyly; originally, Poland's syndrome was named << Poland's syndactyly >>. Finally, we think that we can talk about Poland's syndromewithout anomaly of homolateral upper end, the major element is musculary agenesis of sternocostal pectoralis major. The search of homolateral upper end has to be systematic in front of suspicious of Poland's syndrome.

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## **Poland's syndrome: clinic series and thoraco-mammary reconstruction. Report of 27 cases.**

[Article in French]

Foucras L, Grolleau-Raoux JL, Chavoïn JP.

### **Source**

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## Abstract

The authors evaluate the quality of thoraco-mammary reconstructions in Poland's syndrome. There was 34 patients in our serie, and 27 were operating between 1982 and 2001. There was 19 women and 8 men; the mean age was 19 years (9-40 years). Because of the clinical variability, the authors propose a classification of the malformation's importance in 3 degrees. In our serie, there is 35% of degree I, 53% of degree II and 12% of degree III. Each patient had an average of 2 general anaesthesias (1-6). For the first operation time, men had translation of the homolateral latissimus dorsi muscle flap in 25%, and a thoracic prosthese in 75%. Women had mammary prosthese in 50%, expansion prosthese in 20%, mammary prosthese and thoracic prosthese in 20%, muscle flap in 10%. The same person analysed the results in terms: good, middle or bad. The results are good in 67%, middle in 22%, poor in 11%. In degree 1, the results are always good; in degree II, they are good in 59%, middle in 29% and poor in 12%; in degree III, the results are good, middle and poor in 33%.

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## Latissimus dorsi muscle-musculocutaneous flap in chest-wall reconstruction.

Moelleken BR, Mathes SA, Chang N.

### Source

Department of Surgery, School of Medicine, University of California, San Francisco.

### Abstract

The latissimus dorsi is a versatile muscle that can be employed in a variety of situations requiring chest-wall reconstruction. Its anatomy is predictable and has within it flexibility for transposition to a number of locations based on its standard or reverse arcs of rotation. It can be transposed with a skin island, and its length and bulk can be used effectively to provide durable coverage for anterior or posterior defects. These factors make the latissimus dorsi an excellent choice for the reconstruction of complex chest-wall defects.

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## Reconstruction of congenital chest-wall deformities.

Garcia VF, Seyfer AE, Graeber GM.

### Source

George Washington University School of Medicine, Washington, D.C.

### Abstract

Pectus deformities and Poland's syndrome are two relatively common congenital deformities of the chest wall that are amenable to reconstruction. The extent of the structural deformity in pectus deformity and the degree of associated cardiopulmonary dysfunction are critical variables in preoperative assessment. The operative approaches range from more extensive sternal eversion to the more popular subperichondrial cartilage resection with or without internal fixation. In Poland's syndrome, the options for reconstruction include anterior transfer of the ipsilateral latissimus dorsi muscle through a transaxillary tunnel and attachment to the clavicle and sternum. Submuscular insertion of a mammary prosthesis can be added in the female patient.

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## **Early reconstruction of Poland's syndrome using autologous rib grafts combined with a latissimus muscle flap.**

Haller JA Jr, Colombani PM, Miller D, Manson P.

### **Abstract**

The complex chest wall deformity of Poland's Syndrome may require intervention in early childhood for missing ribs and paradoxical movement of the involved anterior chest with lung herniation. To stabilize the chest wall, we have used autologous rib grafts and prosthetic fabric to replace absent endothoracic fascia. While physiologically sound, this method does nothing for the cosmetic defect which consists of absence of the pectoralis minor and one half to two thirds of the pectoralis major muscles. Silastic implants are not satisfactory in childhood and have complications from recurrent trauma and mobility in young adults. We have had recent experience in three patients (ages 10, 14, and 16 years) with an original technique which results in chest wall stabilization and immediate cosmetic reconstruction without prostheses. The operation utilizes simultaneous rib grafts and a rotational latissimus muscle flap. The muscle not only produces a near-normal contour, but also maintains sensation and contractile function. We propose this technique as a method of choice for the correction of Poland's Syndrome and suggest that it may have wider application in children and teenagers who need soft tissue and muscle replacement of the chest wall for other reasons.

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