POLAND SYNDROME

COMPUTER-AIDED THORACIC CORRECTION WITH CUSTOM-MADE SILICONE IMPLANT

DEFINITION, OBJECTIVES ET PRINCIPLES

Poland syndrome is a relatively rare congenital malformative disorder. Alfred Poland, a student in anatomy, was the first to provide a complete clinical and anatomical description in 1841. The global form of this syndrome associated an agenesis of the sternocostal bundles of the pectoralis major (**Figure 1**) to a malformation of the homolateral hand.

Clinical forms are extremely diversified but the agenesis of the sternocostal bundles is persistent. In women, there is often a mammal asymmetry with a homolateral hypoplasia of the breast and the nipple-areola complex (**Figure 2**).

Poland syndrome is a rare malformation since its incidence is estimated at 1 for 30,000 births. There seem to be a male prevalence (ratio 3:1). Most articles report a right lateralisation of Poland syndrome, with a ratio of 3 for 1.

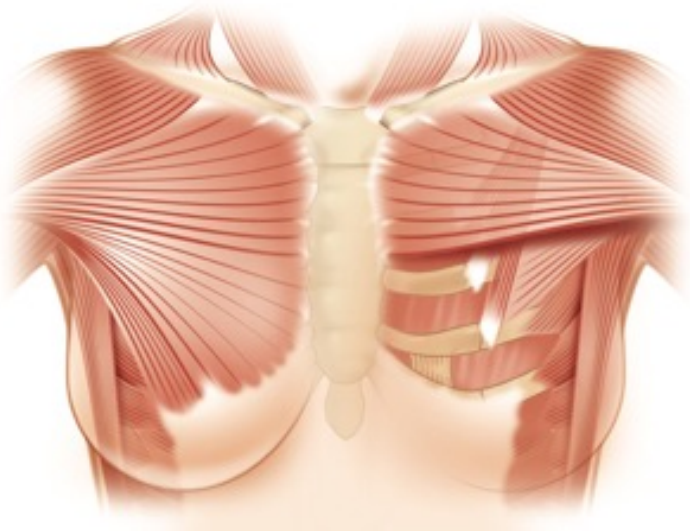


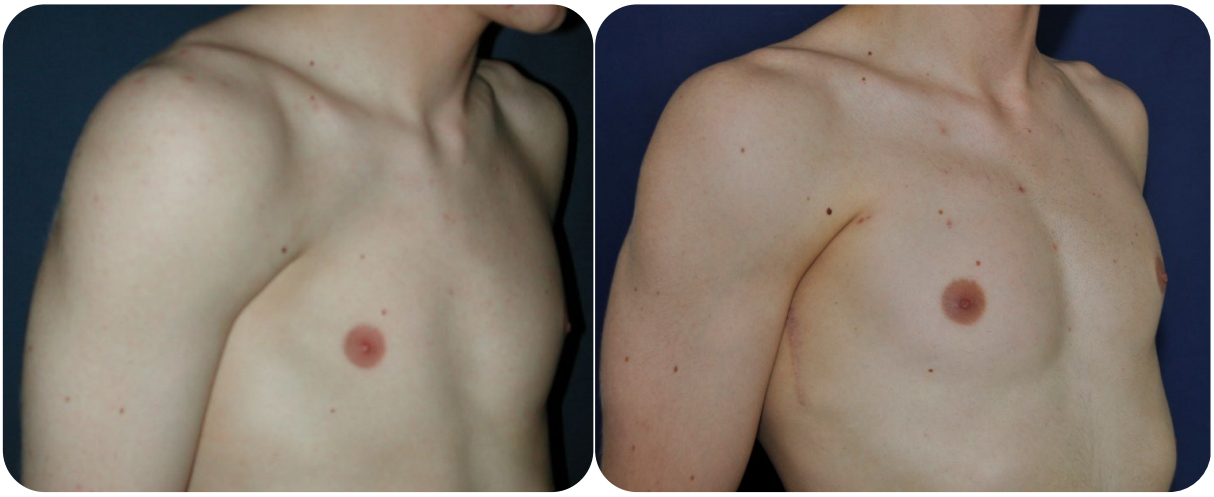
Figure 1

Figure 2

The objective of the reconstructive surgery is simply morphological, with no functional claim, even more so since functional disorders are minor and always well compensated (decrease of muscular strength).

Anatomik Modeling
3D custom-made implants

The principle of the surgery is to increase the volume of the affected thoracic area, using a custom-made implant, in order to compensate the muscular deficiency (**Figure 3-4**). Then, we can consider implementing a breast implant to treat the mammal atrophy in women, and a potential additional transplant of fat tissues for finishing.



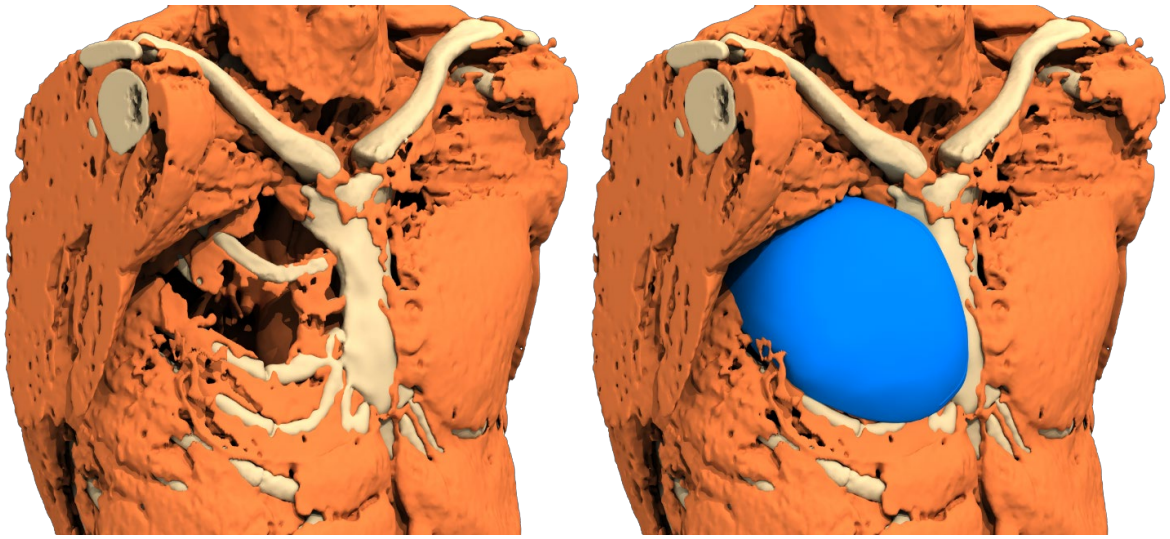


Figure 3

Figure 4

This deformity is often poorly accepted physically and psychologically by the patient, reflected by a modified self-confidence and by a malaise, which can be sometimes deep, leading to a true complex. Psychological consequences are often important from adolescence onwards, disturbing self-image, social relationships, and sometimes indirectly sporting activities.

It is mandatory to wait the end of puberty to perform the intervention, i.e. 14 years old when the hormonal impregnation and the deformity are stable, even if growth is not over yet. If there is a modification of the thoracic morphology due to growth, the implant can be changed later.

The thoracic implants currently used consist of a medical-grade silicone rubber or elastomer: there is no envelope or filling product. The risks of deterioration and breaking are non-existent. It is puncture proof, untearable, and it has an unlimited lifespan. A fibrous exclusion envelope (capsule) is rapidly created, just like around any foreign body (steel, glass, nylon...), but there is never any rejection reaction, producing

antibodies. This envelope cannot retract onto this incompressible implant: there is never any "adhesion" (adhesive capsulitis).

These implants are unique, specific to each patient and made using silicone casting either from a thoracic casting on the patient's skin or increasingly using computer-assisted construction (CAC) from a quality 3D printer (sections from 1 to 1.2mm).

BEFORE THE SURGERY

Patients can be informed of the different existing techniques during a surgical consultation with clinical examination. This consultation is associated to a 3D thoracic scan. Pictures are taken frontally or in 3/4 view.

The 3D thoracic scan must be performed with the arms along the body and in millimetric sections (from 1 to 1.2mm). From this scan, a reconstruction of the patient's virtual body will be computer generated in order to design the virtual implant perfectly suitable to each anatomy (**Figure 5**). This image will be transformed into a resin prototype, which will be used for silicone casting after designing a plaster mould. The medical-grade elastomer implant will eventually be sterilised and delivered to the appropriate surgeon.

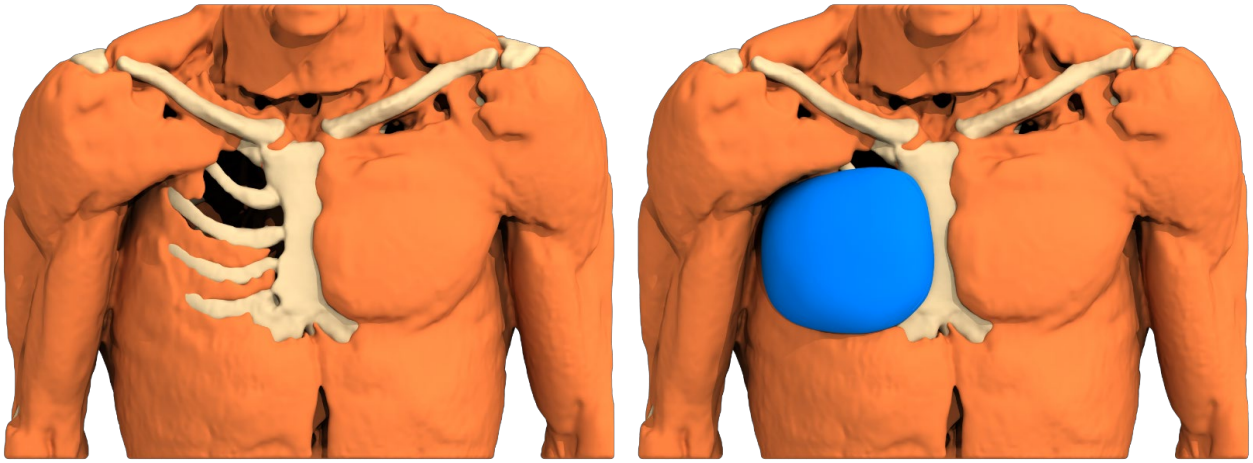


Figure 5: Computer-aided design

TYPE OF ANAESTHESIA AND TERMS OF HOSPITALISATION

The intervention is performed under complete general anaesthesia with intubation in supine position; it requires a 3-day hospitalisation (admission the day before the intervention and discharge the next day or the day after).

THE SURGERY

Preoperative drawing
The surgeon marks the median vertical axis of the thorax, the edges of the implant prototype, and its exact height position (scan reference points).

Skin incision
There is an axillary incision of about 8cm. It continues directly until the costal bony level, preserving the vasculo-nervous pedicle of the serratus muscle.

Detachment
It continues until the edges of the locus drawn on the skin.

Implant insertion
The implant consists of a medical-grade silicone rubber. It is firm in its centre, in the thickest part, but increasingly smooth on the edges, which are tapered like an airplane wing. The folding it thus facilitated as well as the insertion with a minimal incision.

Medical-grade silicone elastomer sterile implant

It is placed in its subcutaneous locus, prepared for its exact size. The implant is perfectly stable and cannot undergo any subsequent displacement, especially downwards.

Closing the wall
It is performed on two levels using absorbable suture material: on the subcutaneous level and using intradermal stitches on the skin.
Thanks to a rigorous haemostasis, aspiration drainage does not need to be performed, which shortens the duration of hospitalisation, the discomfort, and the risk of infection.

Compression
The intervention ends with a dressing and a circulatory compression.

We warn surgeons about the risk of hematoma induced by the use of a suction drain. The strong gradient of depression between the two smooth planes (thorax and implant) can aspirate the coagulation clot of big perforating arteries and cause an early bleeding.

AFTER THE SURGERY: POSTOPERATIVE OUTCOMES

Postoperative pain is most of the time short and controlled with grade 1 analgesics. The absence of muscular section reduces pain compared to other interventions.

A thoracic compression bra needs to be worn night and day for a month.

The sero-haematic and then serous effusion is unstable, due to the oedema in the locus in contact of the implant: it requires a puncture to evacuate from the day of discharge, and then 8 days later.

Work stoppage lasts 15 days, and the recommendation to stop sports is for 3 months; beyond, any sport is possible with no risk or discomfort.

RESULTS

Two to three months are necessary to appreciate the morphological result. It is the time necessary for the oedema to disappear and to improve comfort, which allows for a gradual resumption of sports activities. A whole year is necessary to assess the scarring is good and subtle.

Before/After surgery in a man

Before/After surgery in a woman

Modern computer-aided reconstruction techniques improved the aesthetic results further. Correcting the malformation is in most cases satisfying, but the anatomical restauration of the symmetry is rarely perfect.

POSSIBLE COMPLICATIONS

The correction of a Poland syndrome using a custom-made implant, performed mainly for morphological reasons, remains nevertheless a true surgical intervention, which involves risks associated to any medical procedure, however slight it may be.

Complications associated to the anaesthesia must be distinguished from complications associated to the surgical procedure:

- Regarding the **anaesthesia**, during the mandatory preoperative consultation, the anaesthesiologist will inform himself the patient of the anaesthetic risks. It should be pointed out that the anaesthesia induces reactions in the body which can sometimes be unpredictable and more or less easy to control. However, with a qualified anaesthesiologist-resuscitator, who practises in an actual surgical situation, the risks incurred become statistically very low. It is necessary to bear in mind that techniques, anaesthetics, and monitoring methods have greatly improved over the past thirty years, offering an optimal safety, especially when the intervention is performed outside of an emergency situation and in a healthy individual.
- Regarding the **surgical procedure**, when choosing a qualified and expert plastic surgeon, trained for this type of procedure, you limit the risks as much as possible, without eliminating them entirely though.

In practice, the vast majority of corrections of Poland syndrome using custom-made implants, performed according to the rules, do not present any serious issue; postoperative outcomes are simple and the patients are satisfied with their results even though the symmetry is not perfect. Nevertheless, complications may arise following the intervention.

Complications inherent to the surgical procedure

- **Effusions, infections**

- **Serous effusion**: this is not a complication since it is steady and transient.

- **Haematoma**: the excess of blood around the prosthesis is an early complication which can occur during the first hours. If the haematoma is major, revision surgery in the OR is thus preferable in order to evacuate the blood and to stop the bleeding at its origin. It is very exceptional if all the haemostatic precautions are taken, anticoagulants forbidden, along with trauma, especially an excessive and premature physical activity.

- **Infections**: not described to this day after this type of surgery. A deterrent antibiotic therapy is always prescribed during the intervention, it is not recommended after though.

• **Healing abnormalities**

Since the healing process involves somewhat random phenomena, sometimes scars are not, in the end, as discreet as desired, and might have very different aspects: wide, retractile, adhesive, hyper- or hypopigmented, hypertrophic (swollen), or even exceptionally keloid.
This possibility is rarely problematic due to the axillary position of the scar, which is naturally hidden.

• **Sensitivity modification**

The anaesthesia of the cutaneous area covering the implant is steady but regresses spontaneously in a centripetal manner in a few months.

• **Pneumothorax**

Rare, it requires a specific treatment.

Risks specifically related to custom-made silicone elastomer implants

They are non-existent, instead of flexible silicone gel breast implants.

• No "folding" or "wavy" aspect

• No "adhesion"

• No Rupture

We already stated that implants can be considered as definitive.

• Malposition, displacement

Malposition or secondary displacement of implants is avoided respecting rigorously the surgical technique of installation and the choice of a custom-made computer-assisted conception. It might occur when the implant is implemented very early, sometimes it requires to change the implant.

•No long-term late periprosthetic seroma

Due to the wide clinical variety of Poland syndrome, the therapeutic strategy will be individual, adapted to the degree of severity regarding the malformation, and to the patient's age and gender. The suggested repairing techniques must be safe, with a discreet resulting scarring, since the objective of the correction is aesthetic.

Nowadays, due to progress, it is possible to associate a custom-made thoracic prosthesis, a breast prosthesis or an autologous transplantation of adipose tissue to the demand, depending on the type of malformation. The pedicle flaps of the latissimus dorsi muscle, even the free pedicles and the bone reconstruction have almost no space left in the therapeutic strategy.

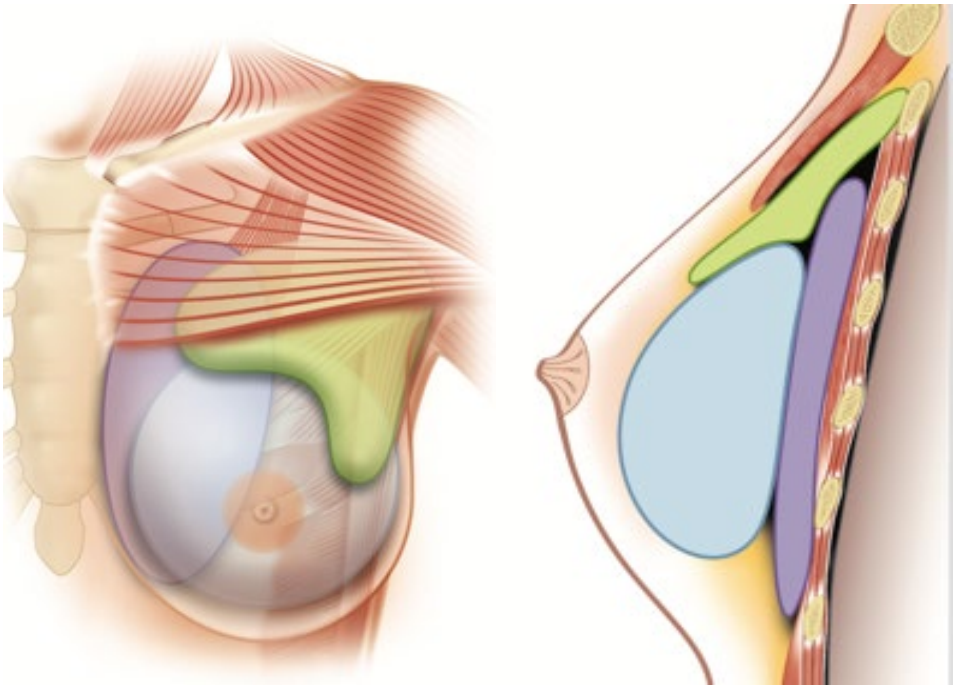


Figure 6: Custom-made thoracic implant (mauve) and additional techniques: breast implant (blue), adipose tissue transplantation (green)

ASSOCIATED BREAST HYPOPLASIA IN WOMEN

In case of asymmetry or associated breast hypoplasia, it is possible to consider implementing secondarily one or two breast implants, but mandatorily after a minimum of 6 months.

A request for prior agreement is mandatory.
The prosthesis may be round or anatomical; it consists of an envelope of silicone elastomer and a filling product. The most used filling product is silicone gel. This envelope may be smooth or textured.

Surgical technique
The intervention is performed under general anaesthesia. The favourite approach is the same axillary approach than for the thoracic implant. Due to the absence of pectoral muscle, the implant can only be placed in a retroglandular position – when there is gland – otherwise it is placed subcutaneously. Thus, the subcutaneous position and the poor quality of the teguments put the patient at significant risk of presenting complications. A first intervention consisting of a transplantation of adipose tissue improves the local trophicity, facilitating the implantation procedure.

Benefits and disadvantages
The main "complication" is a flawed result due to the prosthesis protrusion in the segment I of the breast, which is magnified by the intraclavicular depression which has not been corrected. The other complications are inherent to the implementation of a breast prosthesis.

ADDITIONAL TRANSPLANTATION OF ADIPOSE TISSUE

Coleman's technique, elaborated in 1986, to reinject adipose tissue is used to fill depressions or to restore volumes. It can be performed at the same time as the implementation of a breast implant or later.

Surgical technique

The autologous adipose tissue is withdrawn using a syringe by liposuction. This sample is centrifuged and the intermediary fat phase is reinjected as atraumatically as possible at the site which needs to be filled. This latter is spotted before the surgical intervention, drawing the contour of the atrophic area and is beforehand prepared using an active cannulation to obtain a section of the numerous fibrous tracts, which is sometimes difficult. The adipose tissue is spread with injections in the three dimensions. Graft loss represents between 30 and 50%. Several sessions may be necessary every few months. It improves the infraclavicular depression with a better filling and more modestly helps recreating the anterior axillary pillar.

Benefits and disadvantages

This is a simple technique, which however requires a specialised equipment. The lipofilling technique may be enough for minor forms of Poland syndrome but most of the time it is used with another technique. A limitation of this method is due to poor sites in young and thin donors. The main complications are cytosteatonecrosis and disappointing results. Finally, several sessions are usually necessary. Repeated liposuctions are not trivial (irregularities, cellulite where repeated samples were taken).

CONCLUSION

This is the information we wanted to give you in addition to the consultation. We advise you to keep this document, to read it again after the consultation, and to think about it later.
Then, you may have new questions, for which you will expect additional information. We are at your disposal to talk about it during a next visit, or on the phone, or even the day of the intervention when we see each other again, and of course before the anaesthesia.

Thank you for sending us your **signed and dated written informed consent**, mentioning you agree to the silicone rubber implant, to the 8cm axillary scar, to the general anaesthesia, and to the impossibility of obtaining a perfect symmetry.

PERSONAL NOTES