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CASE REPORT

Lipomodeling of Poland's Syndrome: A New Treatment of the Thoracic Deformity

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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.

Keywords Lipomodeling · Poland's syndrome · Thoracic deformity · Fat grafting · Autologous reconstruction · Breast reconstruction

Poland's syndrome is a rare malformation syndrome of debatable etiology [1–3]. Described for the first time in 1826 by Lallemand [4], the syndrome is named after a student of anatomy who described it in detail in 1841 [5]. Its incidence is underestimated because of the variability of its clinical expression, particularly in the minor forms, but is considered to be about 1 in 30,000 births [6]. The original description of Poland's syndrome includes thoracic involvement with hypoplasia of the pectoralis major muscle and deformity of the ipsilateral hand. However, minor forms occur, with isolated thoracic deformity. Involvement of the upper limb concerns mainly the hand and may present numerous clinical forms. The characteristic deformity of Poland's syndrome is shortening of the middle phalanges of the fingers [6, 7]. In thoracic involvement, the constant abnormality is agenesis of the sternocostal fascicles of the pectoralis major [8]. Other associated thoracic involvement may result in more severe clinical forms. Hypoplasia or even absence of certain muscles of the scapular region and involvement of bone and cartilage may be associated with the pectoral hypoplasia. The involvement of the thoracic structures leads to sternocostal deformities which have little effect on function [8, 9].

In women, the deformity results in asymmetry of the breasts, which often reveals the syndrome at puberty. The breast is abnormal in both size and position, as it is

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generally higher and more external. The nipple-areola complex may also be affected [9].

Until now there has been no technique that could satisfactorily correct the severe forms of Poland's syndrome. The classic techniques required major surgery that left residual scars and required implants that were sometimes poorly tolerated. Moreover, the majority of results obtained by these techniques were inadequate and the appearance was not very natural.

We present a new treatment for a severe form of Poland's syndrome, lipomodeling. Lipomodeling is a technique that we developed initially for breast reconstruction [10–13] but that we applied to treat this patient.

Case Report

When 11 years old our patient sought advice for treatment of Poland's syndrome with marked thoracic deformity of the left side (Fig. 1a, b). She had undergone a surgical procedure when 1 year old for correction of syndactyly of the left hand. On examination, in addition to the signs of syndactyly, she had shortening of the middle phalanges of the fingers. Examination of the breast and thorax showed

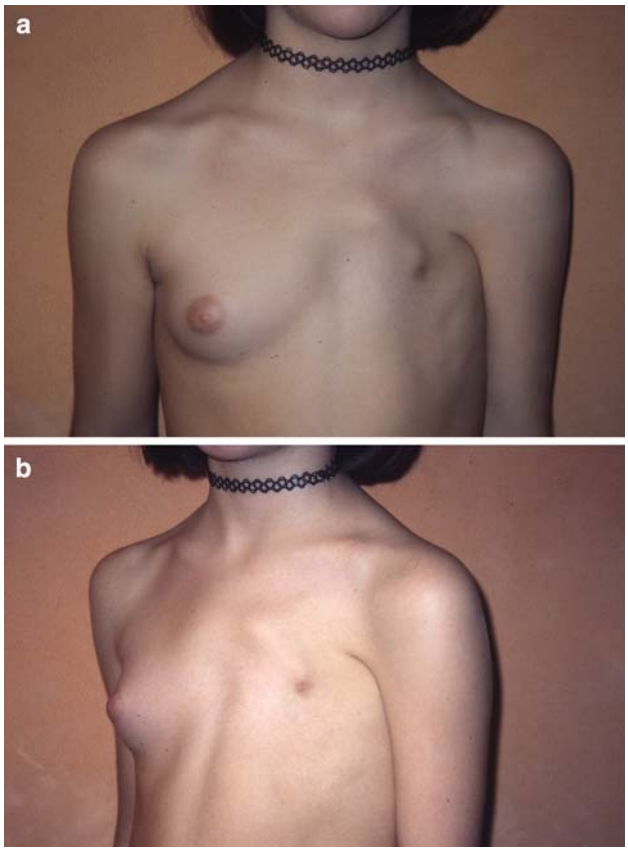

Fig. 1 **a** Preoperative frontal view. **b** Preoperative three-quarter view

Fig. 2 Posterior view showing the lateral depression revealing agenesis of latissimus dorsi

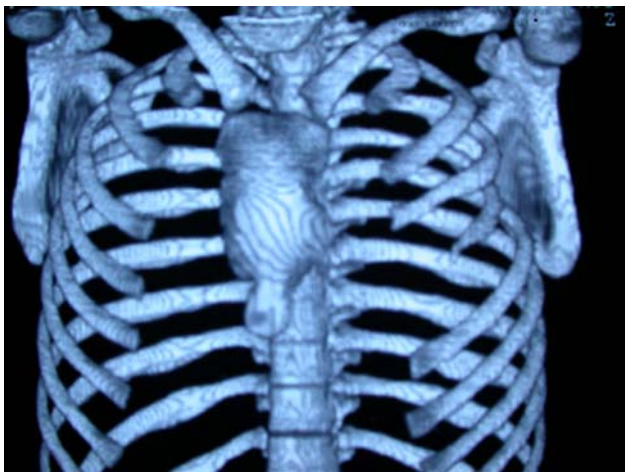

Fig. 3 Three-dimensional reconstruction of thoracic CT scan showing absence of fusion of the 4th rib

agenesis of the pectoralis major muscle and of the latissimus dorsi muscle on the same side (Fig. 2), as well as malposition of the nipple-areola complex. In addition to these soft tissue deformities, there were deformities of the costal arches with absence of fusion of the fourth costal arch (Fig. 3). In this area, only a fibrous and cutaneous tegument covered the lung parenchyma. The patient was 156 cm tall and weighed 43 kg. Treatment was deferred for 1 year to allow the onset of puberty. We thus verified the total agenesis of the mammary gland. Our believed that growth of the normal breast should be accompanied by the surgical growth of the deformed breast, to make it easier for the patient to accept the new breast as an integrated part of her body. The patient and her parents were fully informed about the operative technique, its risks and potential complications, and were given a fact sheet about lipomodeling in breast reconstruction.

Lipomodeling Technique

Lipomodeling was originally developed to augment reconstructed breasts [12]. The procedure is performed with the patient under general anesthesia in the same aseptic conditions as all surgical procedures. The patient is placed in the ventral decubitus position and fat is taken mainly from the thighs and buttocks. A 3-mm harvesting cannula is used without previous infiltration. The incisions are made with a No. 15 blade and concealed in the subgluteal fold. We use 10-cc Luerlock® syringes. In this technique, the operator maintains a vacuum between the aspirate and the plunger in order to reduce damage to the adipocytes. While the fat is being harvested, the theatre nurse prepares the syringes for centrifuging; they are sealed and centrifuged in batches of six for 3 min at 3000 rpm. At the end of the procedure, diluted ropivacaine hydrochloride (Norapin®) is injected via the harvesting cannula to reduce postoperative pain during the first few hours after liposuction. The skin incisions are closed by separate sutures using rapidly absorbed material. A protective adhesive dressing is applied. The patient is then placed in a semi-sitting position for the fat transfer stage. Centrifuging separates purified fat from residues and results in three layers: a top layer containing oil from ruptured fat cells, a bottom layer containing blood residues, and a middle layer containing purified adipocytes. The purified fat is transferred with a 17- or 18-gauge cannula. The skin is incised with the bevel of an 18-gauge trocar so that the scars at the recipient site will measure only 2 mm. Fat is transferred until the recipient zone is saturated (Fig. 4). Rapidly absorbed sutures are used and only simple postoperative care is required. The protective adhesive dressing is left in place for 5 days in the donor areas. The patient leaves the department the following day and is treated with simple analgesics such as paracetamol.

Fig. 4 Perioperative view of the fat injection phase during lipomodeling

Five lipomodeling sessions were performed with a few months between each session, the time necessary for integration into the host tissues and also to fit in with the school timetable of this young schoolgirl. The surgical protocol was the same at each session but fat was taken from different sites (Table 1).

The amounts of fat harvested and transferred after centrifuging were as follows: first session (August 2001), 164 ml harvested and 127 ml transferred; second session (6 months later), 280 ml and 209 ml; third session (4 months later), 150 ml and 82 ml; fourth session, 300 ml and 206 ml; and at the fifth session (one year later), an additional 185 ml was transferred, improving the projection and volume of the reconstructed breast. The nipple-areola complex (NAC) was reconstructed under local anesthesia, the nipple by transfer of the rudimentary ipsilateral NAC which was too high, and the areola by tattooing (Fig. 5a, b).

Postoperative Regimen

The patient was seen regularly after each session. The postoperative course after each procedure was the same as after liposuction, with moderate pain at the donor site relieved by paracetamol, bruising for 2-3 weeks, and edema which resolved in 2-3 months. There was much less discomfort at the recipient site which was practically painless. No particular treatment regimen was prescribed. We merely asked the patient to encourage progressive integration of her breast by practicing sensory rehabilitation, which we recommend after breast reconstruction [14]. The patient is advised to caress the breast daily with the fingertips while, in front of a mirror.

Results

Before the first session we found that the skin seemed to adhere to the fibrous plane of the ribs. Gradual gentle transfer of fat seemed to undermine rudimentary fibrous layers. This first session made it possible to prepare the subsequent transfers. The following sessions were similar to simple lipomodeling of a reconstructed breast, that is, transfer of fat in a three-dimensional network. No surgical complications were observed. Wound healing and revascularization of the fat autografts were satisfactory. There was minor edema in the early postoperative period at the donor sites, but this did not prevent the patient from rapidly resuming her personal and school activities.

The postoperative results were considered very acceptable by the patient, who readily agreed to return for each session. She accepted the new breast as an integrated part of her body particularly well, and each new session was experienced as corresponding to normal breast development.

Table 1 Amount of fat tissue harvested and transferred at each lipomodeling session

Lipomodeling sessions	Months between each lipomodeling session	Harvest site	Amount harvested (ml)	Amount transferred (ml)
20/08/01	0	Buttock thigh	164	127
11/02/02	6	Buttock thigh	280	209
24/06/02	4	Buttock thigh	150	82
16/12/02	6	Abdomen	300	206
08/12/03	12	Buttock	260	185

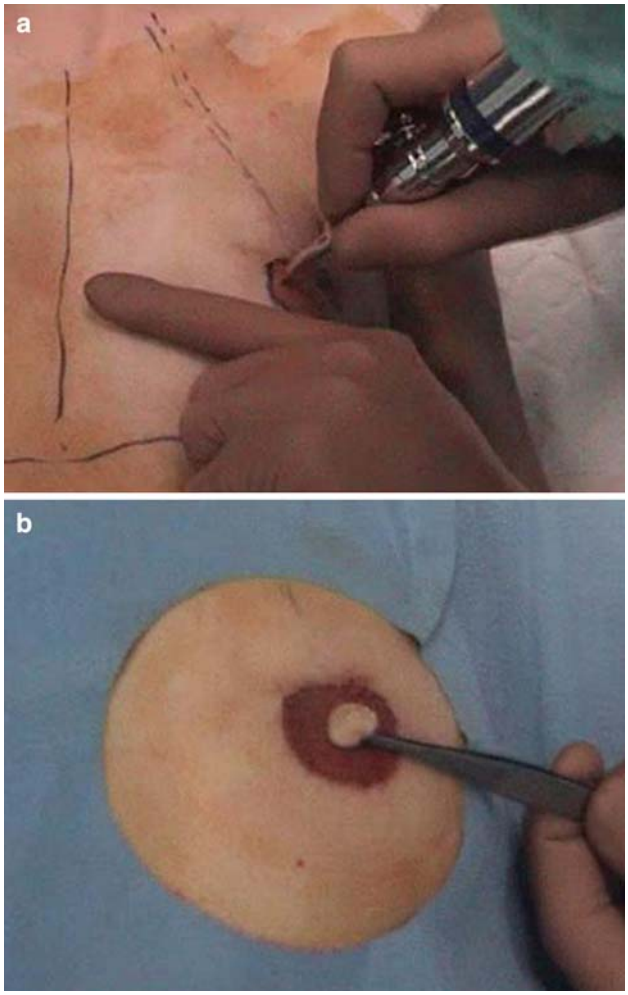

Fig. 5 Reconstruction of the nipple-areola complex. **a** Reconstruction of the areola by tattooing. **b** Reconstruction of the nipple by full-thickness skin graft

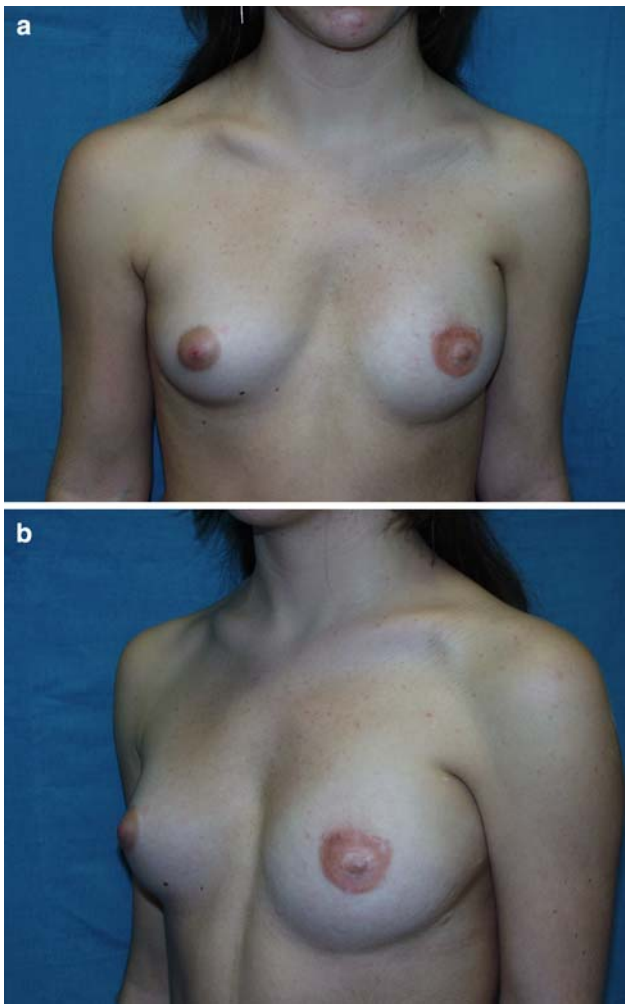

Fig. 6 **a** Postoperative frontal view at 1 year. **b** Postoperative three-quarter view at 1 year

More than 1 year after the last session the result was stable and the breast still had the volume obtained 3 months postoperatively. Detailed evaluation of the result was carried out. The morphological and cosmetic results were assessed by two surgeons, who judged them as excellent. Patient satisfaction was also scored excellent. According to the patient and her parents, they had never hoped to obtain a result of such quality. Breast sensitivity (heat, cold, touch, and light contact in the different quadrants) was considered normal by the patient.

Two-dimensional photographs (Fig. 6a, b) showed impressive improvement, with reconstruction of a breast of practically normal appearance. A three-dimensional study (Fig. 7) using interferometry revealed restoration of breast volume practically identical to the opposite breast, with a slight defect in the projection of the areolar area which is a little flatter and with less projection.

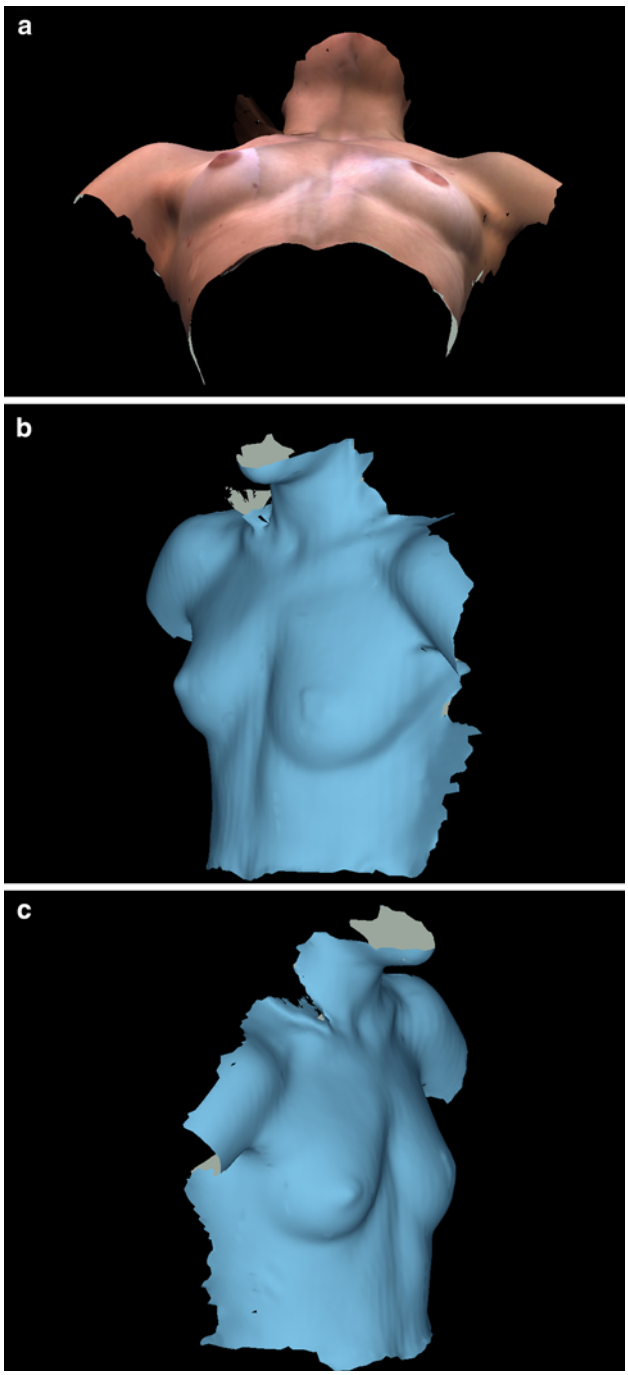

Fig. 7 Postoperative 3D imaging at 1 year. **a** Inferior view. **b** Left three-quarter view without texture. **c** Right three-quarter view without texture

Breast imaging was carried out for reference. Mammography (Fig. 8) showed images that were described by the radiologist as those of a "normal" breast. Echography (Fig. 9a, b) also showed a breast of normal appearance, without cysts or images of cytosteatonecrosis. CT and MRI imaging (Figs. 10, 11) also showed a normal is somewhat fatty breast.

After over 6 years of follow-up, the results of lipomodeling to treat this case of Poland's syndrome are long-lasting and without sequelae at the donor site (Figs. 12, 13).

Discussion

We have confirmed that breast reconstruction using lipomodeling in Poland's syndrome is technically feasible. No surgical complications were observed and the final result is particularly encouraging. The thoracic deformities of Poland's syndrome have been described [8, 9, 15]. Our case corresponds to the most severe forms of the classifications of Shamberger [8] (stage D) and of Glicenstein [9] (stage C). As far as we know, the correction of such a severe case of Poland's syndrome by lipomodeling has never been described and this is the first report in the world literature.

The classic techniques for reconstruction of deformities of the breast and thorax are partly based on those of breast reconstruction [16]. Most of these techniques could not be used in our patient. In particular, transfer of the latissimus dorsi flap, which we use extensively in breast reconstruction, could not be performed in this patient because of the absence of this muscle. Agenesis of the latissimus dorsi [2, 17] has been described in the literature. This anomaly may be associated with hypoplasia of other muscles such as the pectoralis minor, the subclavius muscle, the serratus anterior, intercostal muscles, the rectus abdominis, the external oblique muscle of the abdomen, the deltoid, and trapezius [5, 7, 18–20]. More complex autologous reconstructions such as the transverse rectus abdominis musculocutaneous flap, deep inferior epigastric perforator flap, and gluteus maximus free flap [21] had no place in this case: This young girl, who weighed 43 kg, had no adequate fat deposits for this type of surgical procedure. In addition, she did not want noticeable residual scarring. Reconstruction with an implant or by skin expansion [22–26] would have been difficult because the tissues were very thin and adhered to the rib cage. Also, because of the absence of fusion of the anterior costal arch, the implant could have migrated within the thorax, and to prevent this we would have had to reconstruct the rib cage beforehand. It also would have been necessary to improve coverage of the implant by microsurgical muscle transfer (free contralateral latissimus dorsi, for example) [21] which would have resulted in its own scarring and sequelae at the donor site.

The lipomodeling technique has been developed by our team since 1998. For several years we have been transferring small amounts of fatty tissue in aesthetic surgery of the face, as several authors have shown this to be effective [27–29], including Coleman [27, 30], who comprehensively summed up the surgical principles of preserving the fat and facilitating the take of the grafted adipocytes. The apparent efficacy of the

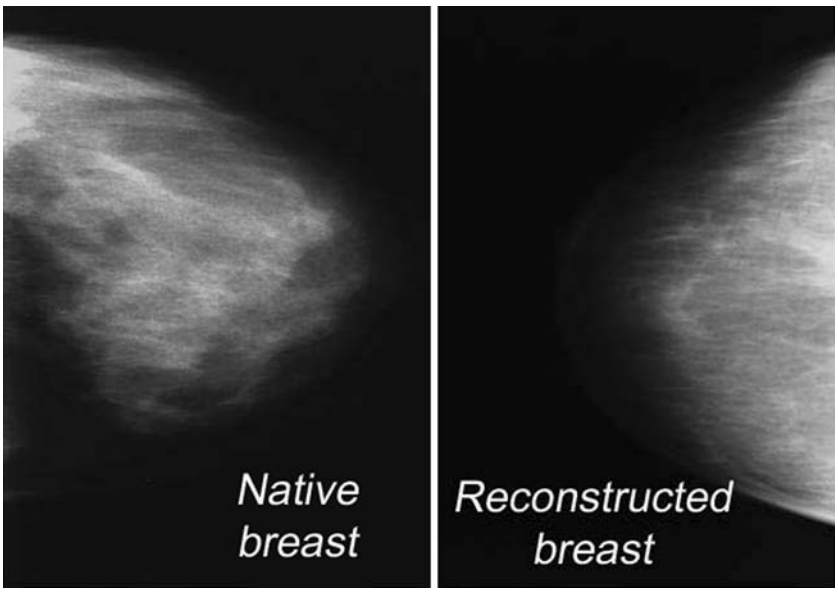

Fig. 8 Mammography of the native and the reconstructed breast

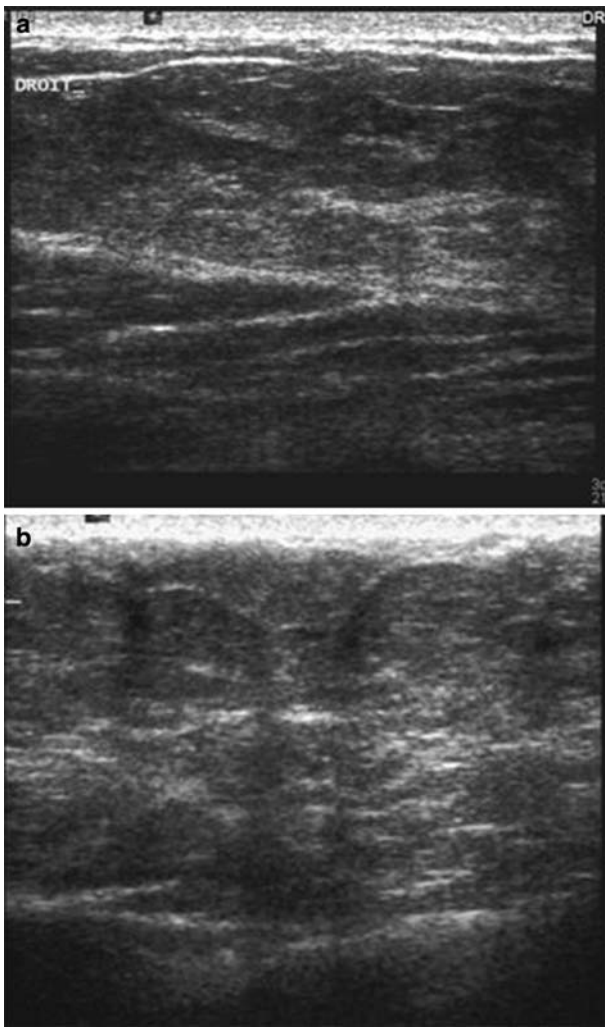

Fig. 9 Postoperative echography at 1 year. **a** Native breast. **b** Reconstructed breast

transfer of small amounts of fat and the need to augment the volume of breasts reconstructed with the autologous latissimus dorsi flap (ALDF), which is inadequate in 30% of cases [10], gave us the idea of using adipose tissue grafts in surgical breast reconstruction. During the second surgical procedure for breast reconstruction, 5 months after reconstruction with an autologous latissimus dorsi flap, patients received a transfer of fatty tissue during the procedure to reconstruct the nipple-areola complex and restore the symmetry of the opposite breast. The technique has been progressively refined. Clinical experience [12] has taught us that large amounts of fat (up to 470 ml per breast and per session in a reconstructed breast) could be transferred, that about 30% of the volume transferred was resorbed during the first 3 months after the procedure, and that there were no complications if a very strict surgical technique was respected [12, 13].

The term lipomodeling (lipomodelage in French) suggested itself to us because of the manual aspect of this procedure: We are in fact modeling the breast with adipose tissue. Adipocyte transfer now makes it possible to consider the autologous latissimus dorsi flap the best choice for autologous breast reconstruction [12, 13]. Lipomodeling enables tailor-made correction of imperfections of the latissimus dorsi flap while retaining the autologous nature of the technique, and it also gives excellent restoration of the décolleté area. The first stage of ALDF reconstruction can be considered as preparation of the recipient site for the lipomodeling stage. The breast reconstructed with ALDF is an ideal recipient site [12], with a survival rate of about 70% for the transferred adipocytes. Lipomodeling of the breast after ALDF reconstruction makes it possible to obtain results that were only dreamed of a few years ago. Breasts of shape, volume, consistency, and mobility close to those of the contralateral breast are now frequently obtained.

Fig. 10 Postoperative CT scan at 1 year

Fig. 11 Postoperative MRI at 1 year of the reconstructed breast. No images of cyst or calcification were found

Fig. 12 Abdominal donor site 6 years after the last procedure

Fig. 13 Buttocks donor site 6 years after the last procedure

Since we could not use a latissimus dorsi flap for our patient and were unlikely to obtain a good result with complex techniques, we decided to use repeated sessions of lipomodeling alone. The advantages of this technique are its autologous nature (without insertion of an implant), low cost, minimal scarring, and absence of sequelae at the donor site. In addition, lipomodeling eliminated the need for prior reconstruction of the missing costal arch. We considered it ethical to propose this technique because it did not rule out any future possibilities and our experience with lipomodeling led us to believe that fat transfer could only be positive. If it was not effective a more complex procedure could still be performed. In fact, repeated lipomodeling alone yielded an almost ideal reconstruction. Only the areolar region showed slightly less projection than that of the opposite breast on close three-dimensional examination by interferometry [31]. Lipomodeling resulted in a breast with a natural appearance and consistency that matched the opposite breast. Fat transfers lead to very few complications [13, 32, 33]. The postoperative course was uneventful, and the patient was able to leave the hospital on the following day with simple analgesics.

The relative drawbacks of this technique are tedious fat harvesting for the surgeon and the need for several sessions. Partial resorption of the fat transferred is another drawback. In our experience with lipomodeling of the autologous latissimus dorsi flap in breast reconstruction, a mean of 30% of the volume injected is resorbed [12, 13]. Therefore, overcorrection during the procedure is necessary. In this case, the recipient site was saturated at each session to make allowances for resorption. There is no doubt that the adipocytes survived, as is demonstrated by persistence of breast volume more than 1 year later, and paraclinical investigations show a breast of fatty type. During reconstruction of the nipple-areola complex, a biopsy of reconstructed breast tissue was

obtained and underwent pathological analysis. The visual appearance and, above all, histological study demonstrated living adipose tissue.

The success of lipomodeling in this very severe case of Poland's syndrome suggests that it will also be effective in less marked clinical forms of the syndrome. We have seven other female patients with less severe forms of Poland's syndrome who are undergoing treatment and they will now be able to benefit from this therapeutic advance. Because we have been able to treat the most severe form of breast and thorax deformity, we have confidence in the lipomodeling technique to treat the less severe forms of Poland's syndrome. However, complementary studies are necessary before final conclusions can be drawn on the indications and contraindications of such a procedure. In particular, it will be interesting to observe how the breasts reconstructed by lipomodeling behave during pregnancies and variations in weight.

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