

# Autologous Reconstruction of a Complex Form of Poland Syndrome Using 2 Abdominal Perforator Free Flaps

*Jaume Masia, MD, PhD,\* Gemma Pons, MD,\* Pietro Loschi, MD,† Lidia Sanchez Porro-Gil, MD,\* Maria Luisa Nardulli, MD,\* and Leyre Olivares, MD\**

**Abstract:** Poland syndrome is the most frequent cause of congenital breast aplasia and hypoplasia. Breast and possible chest wall deformities can be treated with several surgical techniques, including implants, and pedicled or free flaps.

We describe the case of a young patient with severe Poland syndrome with amastia, athelia, and deformity of the chest wall, and aplasia of 2 ribs. Marked hypoplasia of the ipsilateral latissimus dorsi muscle ruled out a reliable reconstructive option.

Two perforator flaps were performed in a single-stage operation. A hemi-deep inferior epigastric perforator flap was harvested to correct the chest deformity, whereas the contralateral superficial inferior epigastric artery flap allowed breast reconstruction.

No complications occurred and a subjectively and objectively pleasing cosmetic result was maintained at 3-year follow-up.

**Key Words:** DIEP flap, SIEA flap, poland syndrome, microsurgical breast reconstruction, abdominal perforator flap

(*Ann Plast Surg* 2013;00: 00–00)

Poland syndrome is a rare congenital deformity (1 in 32,000 live births) with the constant clinical feature of absence of the sternocostal head of the pectoralis major muscle.<sup>1</sup> Hypoplasia of the breast and nipple, aplasia or deformity of the costal cartilages or ribs, and other less frequent findings are described with variable incidence.<sup>2</sup>

The latissimus dorsi (LD) myocutaneous flap, with or without implant, is one of the most frequently used reconstructive options. This muscle is occasionally hypoplastic or absent as part of the syndrome, and therefore not expendable for chest and breast reconstruction.<sup>3</sup> Recently, perforator flaps have been used as a surgical alternative.<sup>4–6</sup>

We present a young female who had a severe form of Poland syndrome with complex thoracic deformity including rib aplasia and almost total absence of the ipsilateral LD muscle. She was treated with 2 simultaneous abdominal perforator free flaps.

## CLINICAL DETAILS

An 18-year-old woman presented with a severe deformity of the right side of the chest wall and lack of solid thoracic support with amastia, athelia, and absence of the pectoralis major pillar (Fig. 1).

Received January 8, 2013, and accepted for publication, after revision, May 3, 2013.

From the \*Hospital de la Santa Creu I Sant Pau, Barcelona, Spain; and †Azienda Ospedaliero-Universitaria di Modena Policlinico, Modena, Italy.

Conflicts of interest and sources of funding: none declared.

Reprints: Pietro Loschi, MD, Azienda Ospedaliero-Universitaria di Modena, Policlinico Del Pozzo St, N.71 41125 Modena, Italy. E-mail: [pietro.loschi@libero.it](mailto:pietro.loschi@libero.it)

Supplemental digital content is available for this article. Direct URL citations appear in the printed text and are provided in the HTML and PDF versions of this article on the journal's Web site ([www.annalsplasticsurgery.com](http://www.annalsplasticsurgery.com)).

Copyright © 2013 by Lippincott Williams & Wilkins

ISSN: 0148-7043/13/0000-0000

DOI: 10.1097/SAP.0b013e31829a39b1

Three-dimensional computed tomography (CT) showed hypoplasia of the third and fourth ribs, aplasia of the anterolateral arch of the fifth and sixth ribs, and reduced volume of the right hemithorax. It also confirmed the absence of pectoralis major and hypoplasia of the LD muscles (Fig. 2) (see Video, Supplemental Digital Content, <http://links.lww.com/SAP/A74>, which demonstrates the preoperative 3-dimensional CT and the CT axial scan showing the chest wall deformity).

Cardiovascular and respiratory functions were normal. No other Poland features were present.

Inventory for the Assessment of Personality Disorders<sup>7</sup> and Symptom Check-List-90 R<sup>8</sup> psychological tests posed no contraindications to surgery (see Video, Supplemental Digital Content, <http://links.lww.com/SAP/A74>, which demonstrates the results of the preoperative psychological tests).

## PREOPERATIVE PLANNING

Treatment of the chest wall depression, providing effective protection for the heart and the lung, and breast reconstruction were offered to the patient. Stabilization of the chest wall with bone grafts or mesh-patch was not necessary as there was no functional impairment.

A bilateral deep inferior epigastric perforator (DIEP) flap was considered the first treatment choice due to the amount of soft tissue required and the need for a large skin island to reconstruct the breast.

CT-angiography (CTA) was used to assess the superficial and deep inferior epigastric systems and their perforators. Patency and caliber of the internal mammary and thoracodorsal systems were judged suitable for anastomosis.

## SURGICAL TECHNIQUE

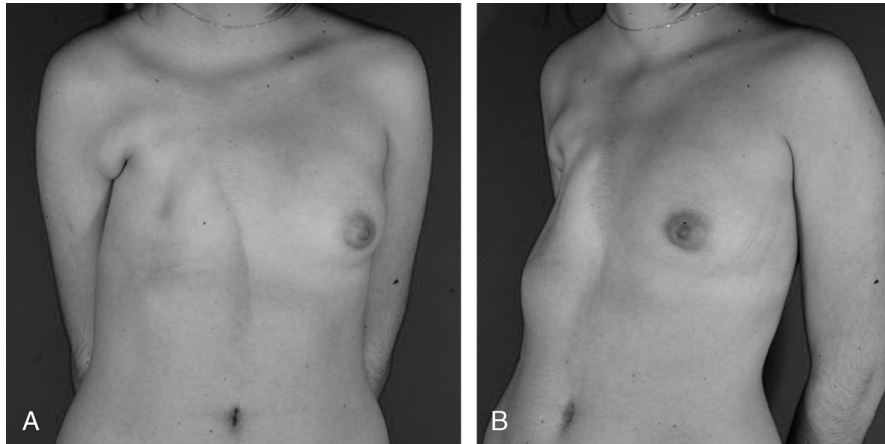
Two hemi-DIEP flaps of 17.5 × 12.4 cm were planned.

On the basis of the CTA information, the superficial inferior epigastric system and DIEPs were preoperatively identified and marked using a handheld Doppler (4 MHz).

A medial row periumbilical perforator was dissected in the left abdomen.

Surgical exploration of the right abdominal wall showed the superficial inferior epigastric artery (SIEA) was competent, whereas the DIEP had a small caliber and was eccentrically placed for possible flap revascularization. Intraoperative assessment of flap perfusion was performed, clamping alternatively the superficial and the deep system, and SIEA dominance was confirmed. We therefore decided to harvest a right SIEA flap.

The internal mammary vessels, displaced more laterally than usual and located subcutaneously due to aplasia of the fifth and sixth anterolateral costal arches, showed good caliber and arterial pulse. No anomaly of the thoracodorsal vessels was detected. After the abdominal flap was separated in the midline, the left DIEP flap was transferred to reconstruct the chest depression and anastomosed to the thoracodorsal vessels (Fig. 3A). The flap was almost completely



**FIGURE 1.** Frontal and left oblique views of an 18-year-old woman with a severe form of Poland syndrome, including right chest wall deformity, amastia, and athelia.

deepithelialized except for a small lateral skin island left for flap monitoring. The SIEA flap was rotated 180 degrees, anastomosed to the internal mammary vessels, and shaped to reconstruct the breast mound. The donor site was closed as for a standard abdominoplasty (Fig. 3B) (see Video, Supplemental Digital Content, <http://links.lww.com/SAP/A74>, which demonstrates the preoperative plan marking, steps involved in the surgical procedure, and immediate postoperative breast and chest wall appearance).

The postoperative course was uneventful and the patient was discharged on the sixth day.

At 6 and 11 months postsurgery, the patient underwent autologous fat grafting to optimize the shape of the flap and treat the contralateral hypoplastic breast (Fig. 4). At 16 months, she underwent nipple reconstruction and areola tattooing.

The patient was highly satisfied with the result (Fig. 5).

## DISCUSSION

Ipsilateral LD myocutaneous pedicled flap is the most commonly used surgical technique for chest soft tissue symmetrization and breast reconstruction in Poland syndrome. Rarely, the muscle may be absent or hypoplastic and unsuitable as a flap for breast reconstruction.<sup>3</sup> Preoperative chest CT or magnetic resonance imaging is therefore mandatory.<sup>9</sup>

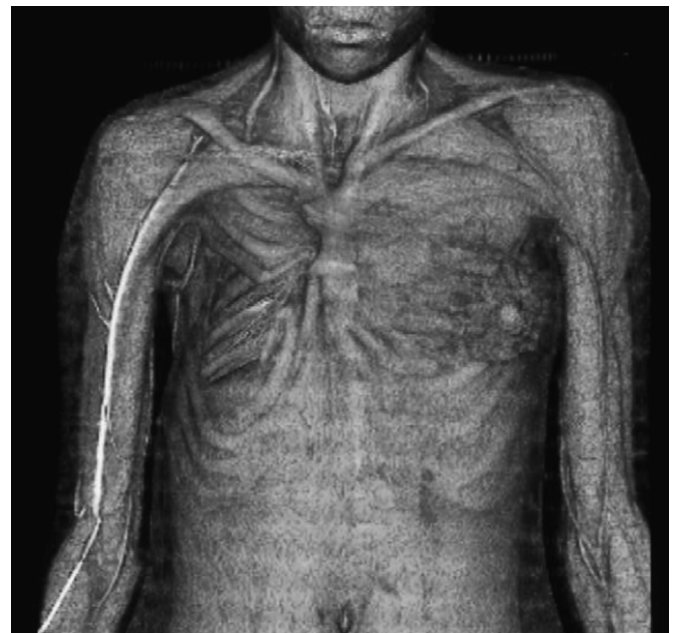
Autologous soft tissue avoids potential complications of standardized breast and custom-made chest wall implants,<sup>10</sup> allowing a more versatile and natural correction of severe and irregularly shaped defects, as described herein.

Interestingly, the first free flap performed for breast hypoplasia, described by Fujino et al<sup>11</sup> in 1975, was planned from the buttock of a woman with Poland syndrome.

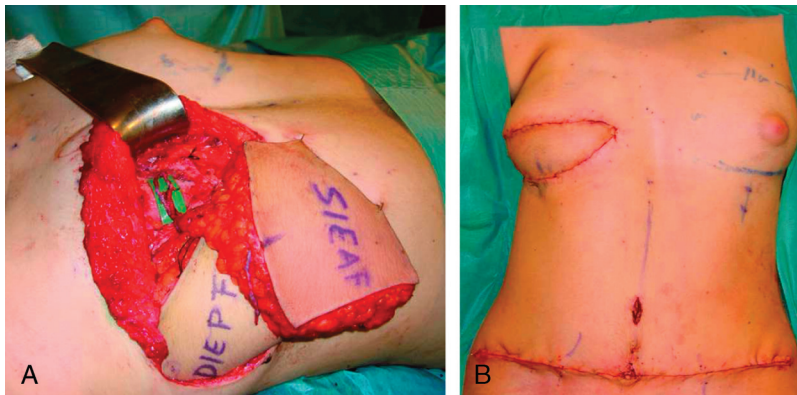
Contralateral microsurgical LD muscles and free transverse rectus abdominis musculocutaneous flaps have been widely described to treat Poland syndrome,<sup>4</sup> although nowadays, perforator flaps are increasingly used in the reconstruction of all sorts of soft tissue defects.<sup>6</sup> Careful assessment of recipient vessels is recommended to detect any anomaly of the internal mammary vessels and subscapular vascular system.<sup>4</sup> Because of the high-quality of abdominal tissue, DIEP flap is considered as the first choice for breast and chest wall reconstruction.<sup>5</sup>

We estimated that a single DIEP flap would not provide sufficient tissue and we therefore planned 2 different hemi-abdominal flaps. To our knowledge, this is the first case of double free flap to correct Poland syndrome chest deformities.

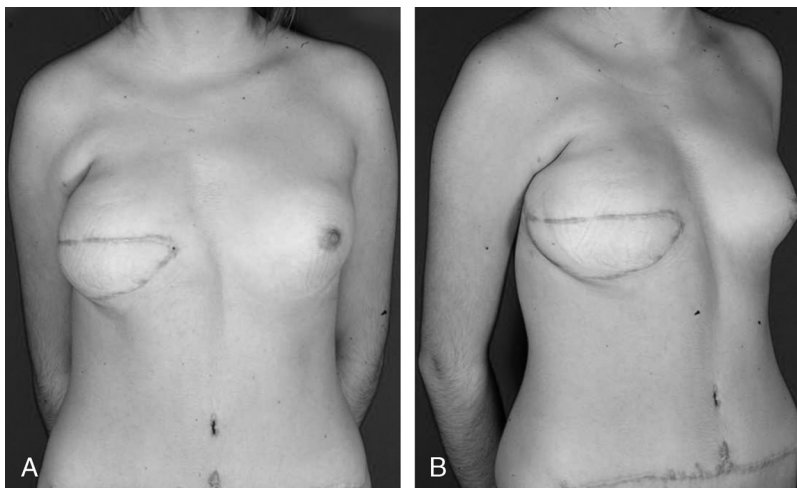
In this case, right abdominal perforators were inadequate for flap revascularization but competent SIEA system allowed to harvest a SIEA flap on one side and a DIEP flap on the contralateral side. These 2 flaps provided an amount of skin and soft tissue that no other pedicled or free flap from buttock or thighs would have provided. The use of 2 flaps also allowed for a better fit and for a more comfortable 3-dimensional reshaping. Both the concealed single scar and minimal donor-site morbidity, associated with a relatively fast recovery time, were additional advantages of the technique. Secondary procedures with bilateral autologous fat grafting were used to refine the contour, volume, and shape of the reconstructed breast to match the contralateral one that was also slightly augmented.



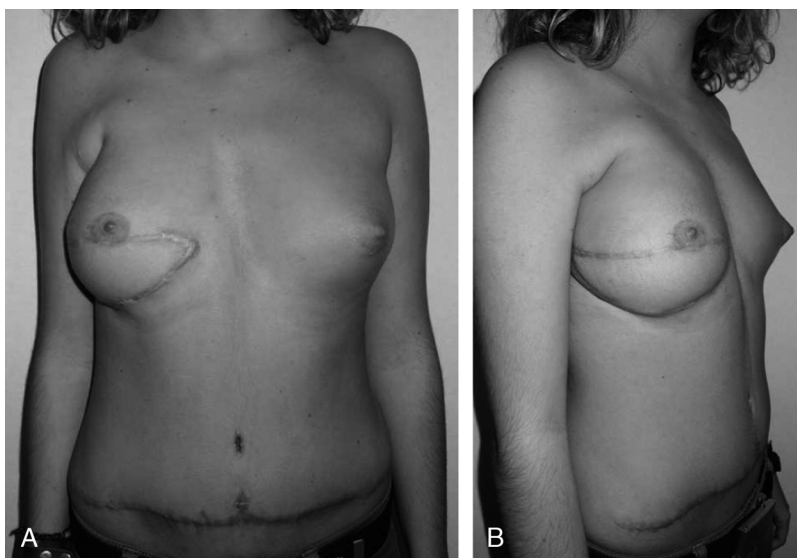
**FIGURE 2.** Three-dimensional CT showing hypoplasia of the third and fourth ribs, aplasia of the anterolateral arch of the fifth and sixth ribs, absence of pectoralis major muscle, amastia, and athelia.



**FIGURE 3.** A, Intraoperative view of DIEP and SIEA flaps insetting. DIEP flap was anastomosed to the thoracodorsal vessels, whereas SIEA flap was anastomosed to the internal mammary vessels, which were located subcutaneously and more lateral than usual (green background). B, Immediate postoperative appearance after flaps shaping and abdominal closure.



**FIGURE 4.** Frontal and right oblique views at 6-months follow-up before fat grafting refinements.



**FIGURE 5.** Frontal and right oblique views at 3-year follow-up after secondary procedures with bilateral autologous fat grafting and nipple areola complex reconstruction. Laser treatment has been suggested for breast and medial abdominal hypertrophic scar.

Assessment of outcome at 3-year follow-up showed an aesthetically pleasing result with good symmetry of the chest, natural appearance of the breast, and absence of abdominal weakness.

#### REFERENCES

1. Poland A. Deficiency of the pectoral muscles. *Guys Hosp Rep*. 1841;6:191–193.
2. Fokin AA, Robicsek F. Poland's syndrome revisited. *Ann Thorac Surg*. 2002;74:2218–2225.
3. Cochran JH Jr, Pauly TJ, Edstrom LE, et al. Hypoplasia of the latissimus dorsi muscle complicating breast reconstruction in Poland's syndrome. *Ann Plast Surg*. 1981;6:402–404.
4. Longaker MT, Glat PM, Colen LB, et al. Reconstruction of breast asymmetry in Poland's chest-wall deformity using microvascular free flaps. *Plast Reconstr Surg*. 1997;99:429–436.
5. Liao HT, Cheng MH, Ulusal BG, et al. Deep inferior epigastric perforator flap for successful simultaneous breast and chest wall reconstruction in a Poland anomaly patient. *Ann Plast Surg*. 2005;55:422–426.
6. Gautam AK, Allen RJ Jr, LoTempio MM, et al. Congenital breast deformity reconstruction using perforator flaps. *Ann Plast Surg*. 2007;58:353–358.
7. Jáuregui Lobera I, Estébanez S, Santiago Fernández M-J, et al. Coping strategies in eating disorders. *Eur Eat Disord Rev*. 2009;17:220–226.
8. Brandizzi M, Fraietta S, Barbieri V, et al. Diagnostic interpretation of depressive symptoms from a psychodynamic point of view. *Clin Ter*. 2010;161:39–44.
9. Bainbridge L-C, Wright A-R, Kanthan R. Computed tomography in the preoperative assessment of Poland's syndrome. *Br J Plast Surg*. 1991;44:604–607.
10. Hochberg J, Ardenghy M, Graeber G-M, et al. Complex reconstruction of the chest wall and breast utilizing a customized silicone implant. *Ann Plast Surg*. 1994;32:524–528.
11. Fujino T, Harashina T, Aoyagi F. Reconstruction for aplasia of the breast and pectoral region by microvascular transfer of a free flap from the buttock. *Plast Reconstr Surg*. 1975;56:178–181.