

Bilateral Free Deep Inferior Epigastric Artery Perforator Flaps for Reconstruction following Mastectomy in Poland Syndrome Patients

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Background: Poland syndrome is classically described as symbrachydactyly, with hypoplasia of the pectoralis major and other upper thoracic musculoskeletal structures. It is thought to be caused by intrauterine interruption in subclavian arterial flow and often includes breast hypoplasia. Affected vasculature can pose a challenge for reconstruction with free flaps because inflow may not be reliable in this patient population.

Methods: We present the rare case of a 28-year-old woman with left-sided Poland syndrome, significant family history of breast cancer, and *BRCA1*+ mutation who underwent bilateral prophylactic nipple-sparing mastectomies with successful immediate bilateral deep inferior epigastric artery perforator free flap reconstruction. The surgical literature in this clinical scenario is also reviewed.

Results: Preoperative computed tomography angiography of the chest successfully demonstrated the patency and quantified the caliber of the internal mammary vessels to support free flap breast reconstruction.

Conclusions: Free tissue transfer is a viable option for breast reconstruction in patients with Poland syndrome undergoing mastectomy guided by preoperative computed tomography angiography to characterize the internal mammary vasculature. (*Plast Reconstr Surg Glob Open* 2023; 11:e5374; doi: 10.1097/GOX.0000000000005374; Published online 6 November 2023.)

INTRODUCTION

Poland syndrome occurs in up to one in 10,000 live births and presents with a spectrum of abnormalities of the thoracic musculoskeletal system and ipsilateral upper extremity. Almost universally, there is unilateral hypoplasia or absence of the pectoralis major.¹ There can also be associated hypoplasia or aplasia of other chest wall structures, including other chest muscles, breast tissue, nipple–areolar complexes, costal cartilages, and ribs.² It is thought to be caused by compromised subclavian artery

blood flow during early fetal development; therefore, it is also referred to as Poland sequence.³ There is a scarcity of literature on free flap reconstruction of the breast in this setting. Here, we present a rare case of successful bilateral immediate deep inferior epigastric artery perforator (DIEP) flap reconstruction after prophylactic mastectomies in a young woman with a significant family history of breast cancer.

MATERIALS AND METHODS

A 28-year-old, White, nonsmoking woman with Poland syndrome and *BRCA1*+, c3748G>T (p.Glu1250) mutation was seen by surgical oncology and plastic surgery with a plan for bilateral prophylactic nipple-sparing mastectomies and immediate DIEP flap reconstruction at the University of Texas Health San Antonio. Age of menarche was 12 years, and the patient had two prior term pregnancies. She had a history of oral contraceptive use for 2 years. Her family history was significant for early onset breast and ovarian cancers with *BRCA1*+ mutation in multiple first, second, and third degree relatives. Her geneticist estimated her lifetime risk of developing breast cancer to be greater than 60%. The patient elected to proceed with

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bilateral prophylactic mastectomies and immediate free flap breast reconstruction.

On preoperative evaluation, she had Fouchras grade I left-sided Poland syndrome with cup size A breasts and more pronounced left hypomastia, and her left anterior axillary fold was less prominent, suggesting absence of the pectoralis major (Fig. 1). Although she had bilateral grade I ptosis, the right breast was more deflated due to prior pregnancy with a tighter left breast envelope due to her syndrome. She had no upper extremity manifestations of the syndrome. Magnetic resonance imaging verified ipsilateral absence of the pectoralis major and excluded any breast or axillary lesions. Preoperative computed tomography angiography (CTA) of the abdomen verified appropriate DIEP perforator anatomy bilaterally, whereas CTA of the chest demonstrated patent internal mammary arteries bilaterally (2.5 mm on the right and 2 mm on the left at the level of the third rib; Fig. 2).

RESULTS

The patient underwent bilateral prophylactic nipple-sparing mastectomies by the surgical oncology team via inframammary fold incisions. Absence of the left pectoralis major muscle with a normal pectoralis minor was evident after completion of mastectomies. Immediate bilateral DIEP free flap reconstruction was performed by the plastic surgery team. The fasciocutaneous flaps were harvested as true muscle- and fascia-sparing flaps each with two perforators. The internal mammary vessels were exposed on each side via a rib excision approach. The arterial vessel sizes matched preoperative imaging findings. Arterial anastomosis for each flap was performed in an end-to-end hand-sewn fashion, and the venous anastomoses were end-to-end using 2-mm couplers. Postoperative monitoring was performed in the ICU setting by clinical assessment of externalized skin paddles in the lower poles. University Hospital is a training institution with personnel for postoperative monitoring that includes residents and nursing staff who are not part

Takeaways

Question: In addition to preoperative computed tomography angiography (CTA) of the abdomen, what other imaging modality is advisable before free flap reconstruction in patients with Poland syndrome undergoing mastectomy?

Findings: CTA of the chest can delineate the anatomy of the internal mammary artery, confirming patency and quantifying the caliber of vessels in Poland syndrome patients as appropriate recipients for free flap breast reconstruction.

Meaning: This article suggests that in addition to abdominal CTA before free tissue transfer, chest CTA can be used to identify appropriate surgical candidates in the Poland syndrome patient population.

of a dedicated flap monitoring unit. Implantable Doppler probes were therefore used for all four anastomoses, as our junior trainees and nursing staff may not be clinically experienced to detect subtle signs of early flap compromise seen with conventional monitoring alone or internal Doppler monitoring of the venous system only.

The patient's postoperative course was complicated by a nonthreatening hematoma of the right-sided reconstruction that was treated with incision and drainage. At a second stage, she underwent revision surgery to bury the skin paddles in addition to low volume fat grafting to the upper poles, thereby correcting stargazer and step off deformities and restoring her preoperative appearance. At 3 months, she was very satisfied with her outcome (Fig. 3).

DISCUSSION

According to the American Cancer Society, 5%–10% of breast cancer cases are thought to be hereditary, with 50% of these associated with BRCA 1 or 2 genetic mutations.⁴ Due to the high risk of developing breast cancer, it

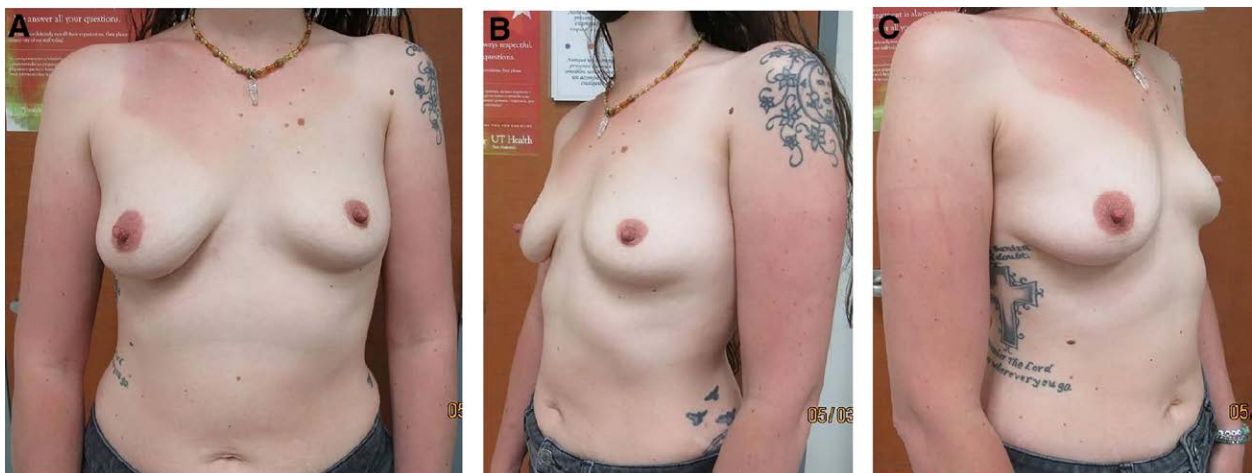


Fig. 1. Case study patient. Preoperative photographs demonstrating syndromic effects on the left breast (A) and blunted prominence of the anterior axillary fold on the left (B) compared with the right (C).

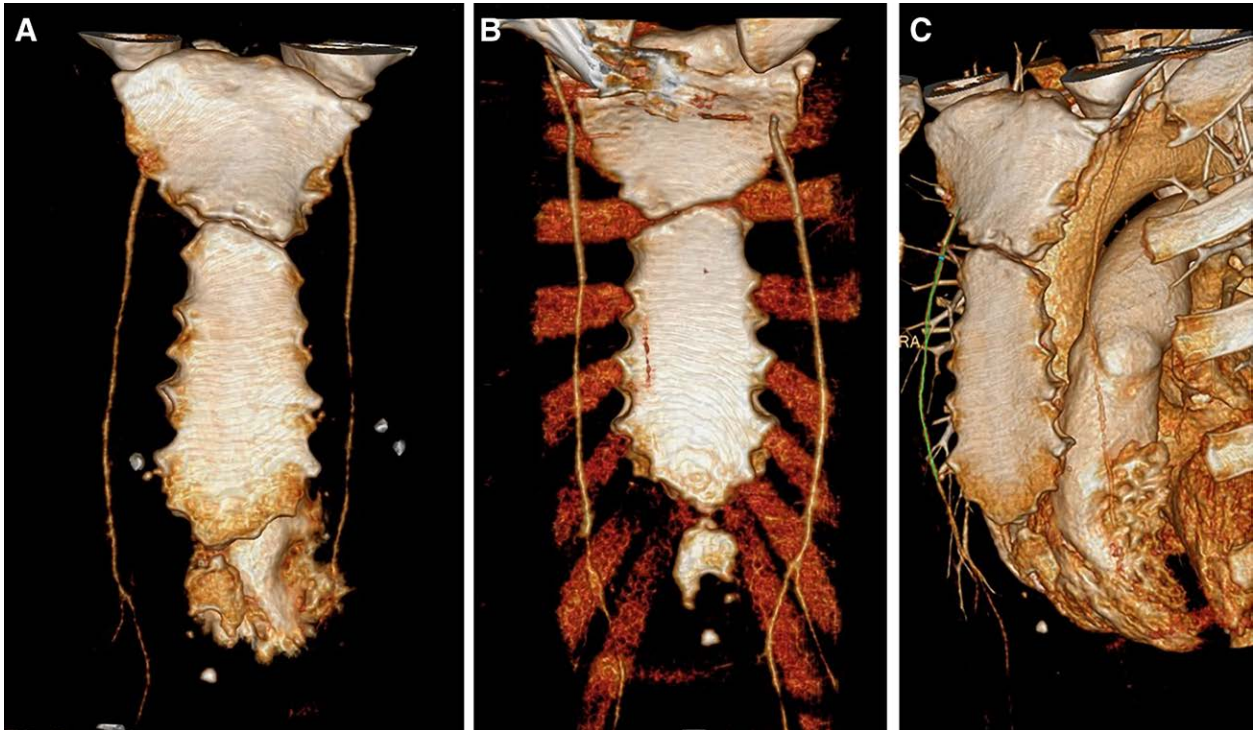


Fig. 2. Three-dimensional reconstitution of preoperative chest CTA demonstrating patent internal mammary arterial vessels bilaterally and favorable vascular architecture. A, Anterior view. B, Posterior view. C, Left oblique view.

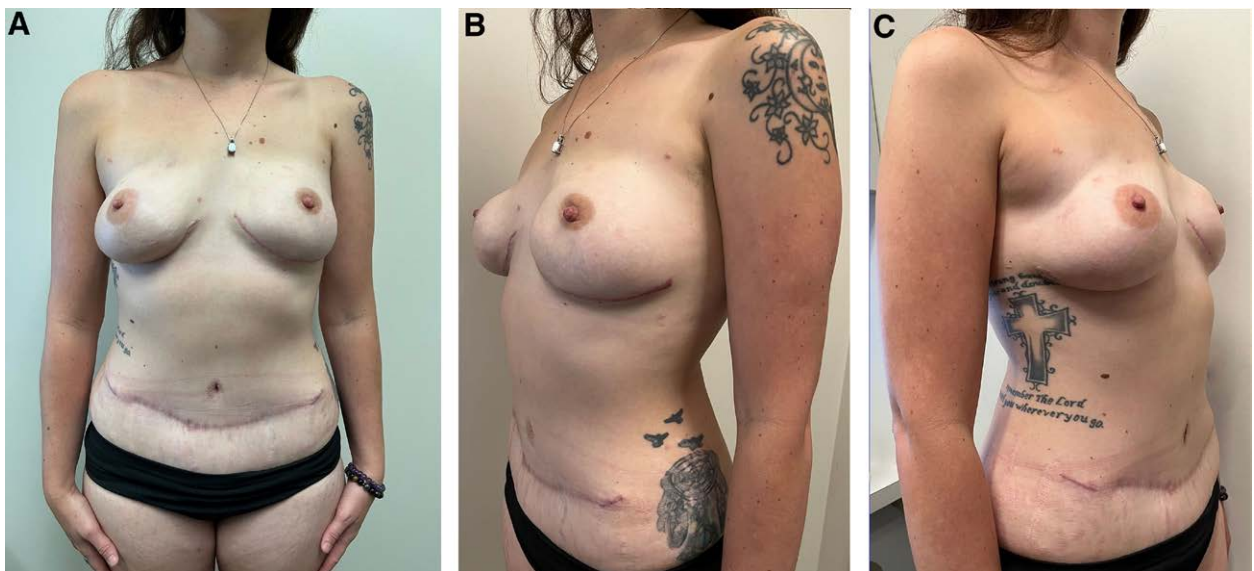


Fig. 3. Case study patient. Three-month postoperative photographs (A–C) showing the results of bilateral free flap reconstruction after nipple-sparing mastectomy and second stage revisions to bury the skin paddles with upper pole fat grafting.

is generally recommended that patients with BRCA positivity undergo risk reducing surgery with bilateral prophylactic mastectomies before the onset of disease. The result of this treatment strategy is that many young women must decide on the best options for postmastectomy reconstruction, which may include no reconstruction with or without use of an external breast mound prosthetic, one

or two-stage implant-based reconstruction, autologous reconstruction, and implant-autologous reconstruction.⁵ Acellular dermal matrix products may be used in either the prepectoral or subpectoral plane to serve as an internal scaffold to enhance the outcomes of implant-based reconstruction, which now represents 80% of breast cancer reconstruction in the United States.⁶ Autologous

reconstruction is a viable option for all patients who are fit candidates for the surgery with firmer indication in the setting of prior adjuvant radiation therapy and prior poor wound healing or infections at the mastectomy site associated with implant loss.⁵

To our knowledge, this is the first reported case of successful bilateral immediate DIEP flap reconstruction after prophylactic nipple-sparing mastectomy in a patient with Poland syndrome and *BRCA1*+ mutation. We elected for DIEP flap reconstruction due to the increased patient satisfaction demonstrated in several high-quality publications comparing use of autologous tissue to implant-based reconstruction in long-term follow-up.⁷ Our patient is young, and we expect that over her lifetime of several decades, this reconstruction would afford her those long-term benefits in terms of appearance; softness; better symmetry; reduced implant-related complications, such as capsular contracture and rupture; and fewer additional operations to address these problems. Finally, this well-informed patient requested a total-autologous reconstruction, as she preferred to avoid implant placement in fear of the risk of foreign material placed in her body and anaplastic large cell lymphoma.

In the first stage of DIEP flap surgery, we prefer to expose skin paddles to facilitate conventional monitoring. Clinical monitoring of the skin paddles added to the use of internal Doppler probes provides several monitoring modalities for detecting flap compromise and making rapid decisions. Although reports have demonstrated earlier time to detection of flap compromise, to our knowledge, no study has demonstrated that use of implantable Doppler monitoring reduces the time for detection of arterial or venous thrombosis as a specific outcome measure compared with clinical monitoring alone. Although the literature varies on this point, several prior studies, including a recent meta-analysis, have reported the more general benefits with the addition of implantable Doppler monitoring to clinical monitoring, including improved time for detection of flap compromise and time for re-exploration, as well as flap salvage and overall flap survival rates.⁸⁻¹⁰

In a previous report of a 51-year-old woman with Poland syndrome and multifocal ductal carcinoma in situ, the patient was successfully treated with a unilateral skin-sparing mastectomy and DIEP flap reconstruction.¹¹ A prior study reported immediate reconstruction with a pedicled transversus rectus abdominis myocutaneous flap in a 42-year-old patient with Poland syndrome and invasive ductal carcinoma.¹² In the majority of other cases of Poland syndrome with a breast tumor, reported reconstruction after mastectomy was predominantly implant-based and less commonly by use of pedicled flap options.¹³⁻¹⁷

Although there is a paucity of studies on free flap reconstruction in Poland patients undergoing mastectomy, there are more reports on treatment of the nonneoplastic breast and chest wall abnormalities related to the syndrome. In an earlier report, listed flaps included free transversus rectus abdominis myocutaneous flaps, superior gluteal artery flaps, inferior gluteal artery flaps, and latissimus dorsi myocutaneous flaps.¹⁸ For the free flaps, recipient vessels reported were branches of the subscapular

vascular axis, which are not dependent on the subclavian system that can be affected in Poland syndrome.¹⁹

Lymperopoulos et al reported DIEP flap reconstruction in three patients with Poland syndrome as a salvage operation.²⁰ All three patients had prior implant-based reconstruction but were not satisfied with the outcome due to capsular contracture in two and implant rupture in the third. These authors performed preoperative CTA to elucidate the anatomy for both the DIEP system and the internal mammary vascular system. Due to the inclusion of younger patients with inadequate abdominal donor site volume in this study as well as in a similar work by Mahrhofer, the authors suggested that free DIEP flaps may not be a suitable reconstructive choice in a significant number of patients.^{20,21} Alternative free flap donor sites, such as from the thigh or gluteal region, may be more suitable depending on the patient's needs.

The inciting event in Poland syndrome is thought to be intrauterine vascular interruption leading to downstream abnormalities of the systems dependent on the subclavian arterial axis. Although it is common practice to radiographically elucidate the abdominal-based vascular anatomy as part of DIEP flap preoperative planning, it is not common to do so for the internal mammary vessels that would be used to provide inflow for the flaps. This investigation is important to perform in patients at risk for congenital vascular abnormalities in the chest, including those with Poland syndrome. Once favorable vascular anatomy is confirmed radiographically, the efficacy and safety profile of free DIEP flap reconstruction in Poland syndrome patients can be made equivalent to the general population.

Liao et al reported successful DIEP reconstruction in a 52-year-old woman with right-sided Poland syndrome that included right-sided breast hypoplasia.²² The authors used preoperative Doppler ultrasound to confirm favorable anatomy of the internal mammary artery and vein to support the free flap anastomosis. In our study, we used preoperative CTA, which provides more information than just vessel size and patency. Vessel architecture is better delineated, leading to more detailed preoperative planning.

One of the largest series of free perforator flap reconstruction for patients with Poland syndrome without mastectomy was by Gautam et al.²³ The authors reported 12 unilateral free flap reconstructions anastomosed to the internal mammary system over an 11-year period with no flap losses but varied aesthetic outcomes. In this study, preoperative imaging was not described, and suitability of the recipient vessels was determined by surgical exploration at the time of flap reconstruction. Given the current widespread availability of CTA technology at relatively low cost, preoperative imaging would seem to be a reasonable option before proceeding with these cases.

Lastly, it is well documented that long-term outcomes from free DIEP flap reconstruction after mastectomy is associated with a high level of patient satisfaction compared with implant-based options.^{24,25} Comparative reports on quality-of-life outcomes for patients with Poland syndrome undergoing various flap reconstructions are scarce,

and none have investigated outcomes after mastectomy in this patient population.²⁶

CONCLUSIONS

In Poland syndrome, hypoplasia of the subclavian-dependent internal mammary system may preclude its

use in free flap reconstruction of mastectomy defects. For these patients, preoperative CTA of the chest can verify the status of the internal mammary vessels to support DIEP flap reconstruction. Primary use of the internal mammary system when favorable preserves the thoracodorsal system, and in the event of free flap failure, it would be available for flap salvage in emergent takeback surgery. If the free

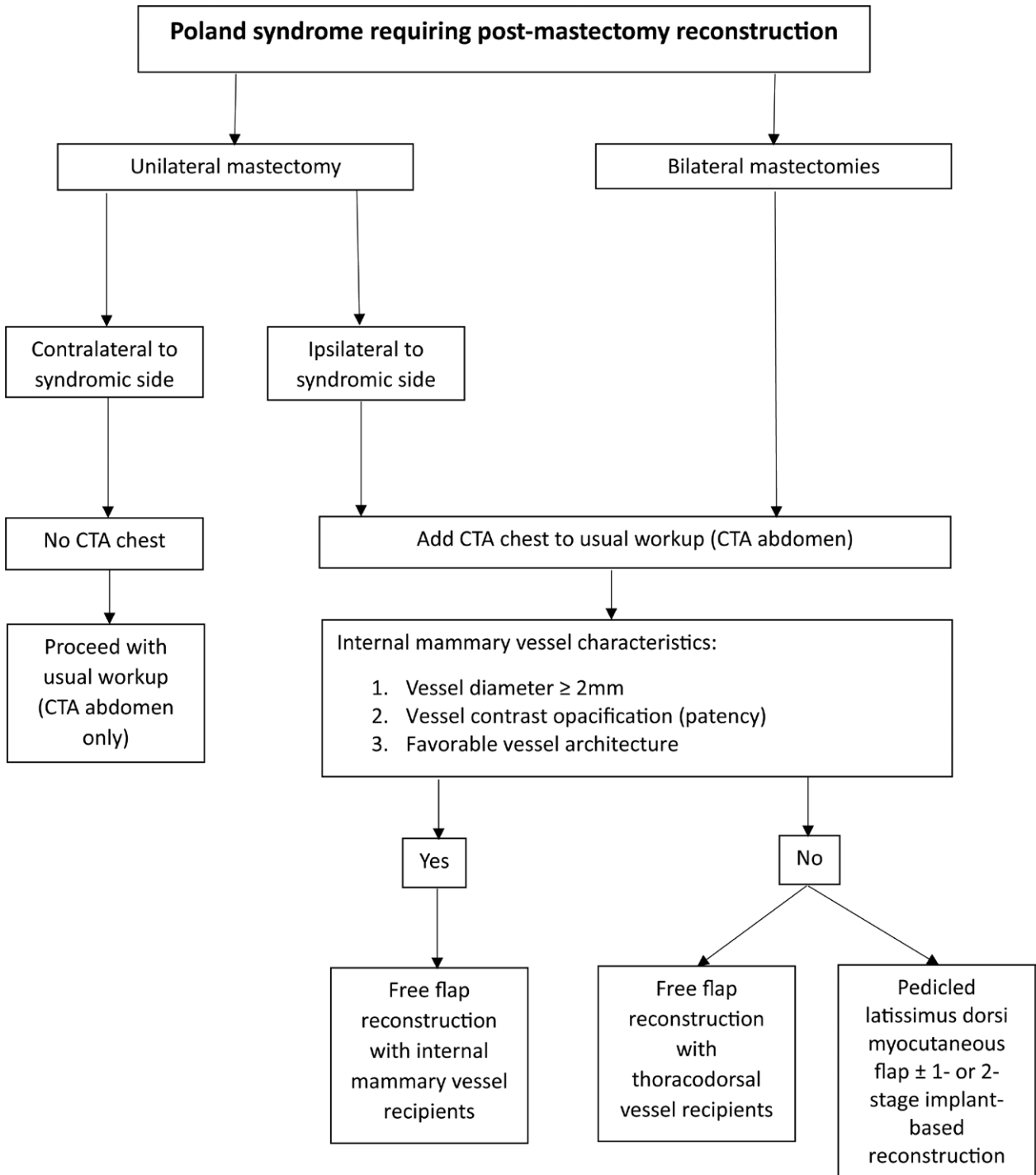


Fig. 4. Clinical flowchart of the preoperative chest imaging workup for Poland syndrome patients requiring breast reconstruction for mastectomy defects.

flap cannot be salvaged and the thoracodorsal system were not used, a preserved thoracodorsal system allows an ipsilateral pedicled latissimus myocutaneous flap to be used to reconstruct the deficient anterior axillary fold and breast along with one- or two-stage implant-based reconstruction if needed to provide additional breast volume.

If the imaging demonstrates unfavorable internal mammary vessels, the surgeon can avoid unnecessary exploration in the chest, potentially resulting in fewer incisions and surgical scars and quicker surgical recovery. A directed surgical approach to alternate vessels such as the thoracodorsal system or use of an alternative reconstruction such as a pedicled latissimus dorsi myocutaneous flap with one- or two-stage implant-based reconstruction can be used.

In the past, preoperative imaging transformed the approach to DIEP flap reconstruction with abdominal CTA and mandibular reconstruction with virtual surgical planning using CTA.^{27,28} This would also facilitate more recent goals for DIEP reconstruction in the general population toward Poland syndrome patients for enhanced recovery after surgery and discharge planning rules by the Centers for Medicare & Medicaid Services that mandate shorter time to discharge after DIEP flap surgery while providing a reconstruction associated with high long-term patient satisfaction.^{7,29,30} We propose that a paradigm shift in the workup of Poland syndrome patients undergoing free flap reconstruction with preoperative CTA of the chest would be of benefit to this population (Fig. 4).

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DISCLOSURE

The authors have no financial interest to declare in relation to the content of this article.

PATIENT CONSENT

The patient provided written consent for the use of her image.

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