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Gigantomastia

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The idea of beauty and "normality" of breast has undergone many changes over the years, depending upon customs and society. To date, breasts are considered normals when symmetric, with a volume ranging between 250 and 400 ml, and the nipple-areola complex situated above the inframammary fold.

Morphologic differences exist between different races, that also depend on weight, age, height and thoracic structure of the patient. Therefore, it is rather hard to establish universal anatomic and clinic criteria to mark a clear-cut limit between normality and hypertrophy of the mammary glands.

However clinical considerations permit to evaluate the presence and gradation of hypertrophy in a particular patient. In this regard, clinical important features are:

- Breast volume;
- The distance between the middle point of the clavicle and the nipple-areola complex;
- The distance between the inframammary fold and the nipple-areola complex.

In one of the most accepted classifications, considering standard breast volume as ranging between 250 and 400 ml, hypertrophy is defined mild for volumes between 400 and 600 ml, moderate between 600 and 800 ml, severe between 800 and 1000, gigantomastia over 1000 ml (Figure 19-1).

A distance of 16 to 21 cm between the mid-clavicular point and the nipple-areola complex is considered "normal", but this value is considerably influenced by patient height. Distance between the nipple-areola complex and the inframammary fold is usually 5-8 cm.

Macromastia or mammary hypertrophy is a deforming, disabling and painful condition characterized by an enlargement of various degree of one or both breasts. Besides being a significant aesthetic defect, this condition causes physical and psychological problems.

Clinical manifestations associated to mammary hypertrophy are:

- Intertriginous lesions induced by friction of the breast against thorax and by the bra at the level of the shoulders; these lesions are worsen by perspiration, which predisposes to infections by Candida, increasing irritation of reddened areas;
- Spinal deformity, with progressive kypho-scoliosis or dorsal kyphosis with attenuation of lumbar lordosis;
- Deformity of sternum-clavicular bony structures, with subsequent headache, neck and shoulder pain, hand paresthesia and breath difficulties;
- Sleep disorders, difficulty in dressing and accomplishing certain movements.

An ulnar neuropathy has also been described in women with severe breast hypertrophy, who report paresthesia in the ulnar nerve territory.

Moreover, psychological problems can negatively influence social and sexual life. Reduction mammaplasty has to be considered, in such a clinical picture, the best therapeutic approach for these patients.

NORMAL ANATOMY

The breast is a pair and symmetric skin relief situated on the anterior surface of the thorax, between the third and the seventh rib, extending from the parasternal line to the middle axillary line. The nipple should lie above the inframammary fold and is usually level with the fourth rib.

The mammary gland, being derived from the ectoderm, is contained in the superficial layer of the subcutaneous tissue, between the superficial fascia and the skin. It is anchored to the pectoralis major fascia by the suspensory ligaments first described by Cooper in 1840, which run from the deep fascia throughout the parenchyma to attach to the dermis of the skin. An horizontal fibrous septum originates from the pectoral fascia along the level of the fifth rib, dividing the mammary gland in a cranial and a caudal part. It acts as a suspensory system, and as a guiding structure for vascular and nerve supply.

Vascular anatomy

The blood supply to the breast relies on two main pedicles: the superolateral pedicle of the external mammary artery (branch of the lateral thoracic artery) and the glandular

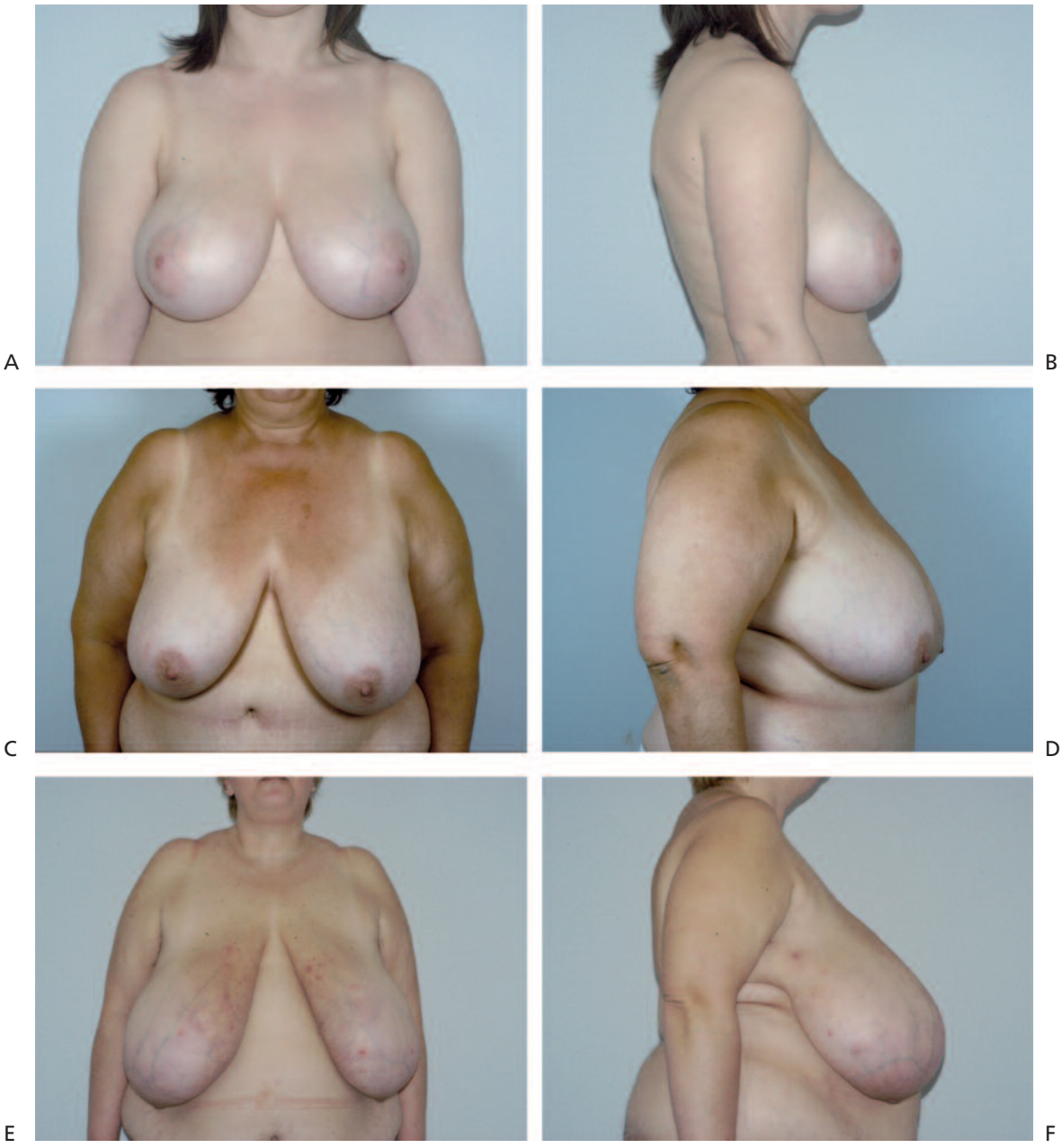

Fig. 19-1. *A-B,* Moderate breast hypertrophy. *C-D,* Severe breast hypertrophy. E-F: Gigantomastia.

branches of the thoracodorsal artery, and the internal pedicle of the internal mammary artery, with its perforators from the second to sixth interspace. Their superficial branches anastomose in the subdermal plexus, supplying the breast skin.

A large perforator of the internal mammary artery emerges from the second or third interspace, running about 1 cm deep to the skin, and supplying a superior pedicle for the nipple-areola complex. Perforators from the third to sixth interspace also present a superficial course, and supply a medially based pedicle. The lateral pedicle relies on the lateral thoracic system, that is usually found 2 or 3 cm deep to the skin at the level of the inframammary fold.

Innervation

Sensory innervation of the breast is mainly derived from the anterolateral and anteromedial branches of thoracic intercostal nerves T3-T5.

Innervation of the nipple-areola complex depends on the lateral branch of the fourth intercostal nerve, with its superficial and deep branch. The first one supplies a lateral pedicle, while the deep branch can sometimes be preserved with an inferior or central pedicle. The anterior branch of the third intercostal nerve also contributes to the sensitivity of the nipple-areola complex; it takes a superficial course within the subcutaneous tissue and terminates at the medial areolar border.

PATHOGENESIS

The breast is the target organ for numerous hormones, which are responsible for mammogenesis, lactogenesis and galactogenesis.

Some authors claim that alterations in endocrine arrangement concerning estrogens, progesterone, prolactin LH, FSH, could represent the "primum movens" of mammary hypertrophy. Others attribute to those hormones only a causative role.

Endogenous hormone stimulation seems to play an important role in juvenile and pregnancy-induced mammary hypertrophy, in which a rapid and massive enlargement of the breasts occur. Subsequent pregnancies are likely to cause further recurrence, once pregnancy-induced mammary hypertrophy has occurred.

Yet in the majority of patients, it is not possible to identify a precipitating cause.

The main pathogenetic hypothesis of severe juvenile and pregnancy-induced mammary hypertrophy are:

- An increase in serum level of estrogens, prolactin, prostegerone, HGC, HLP, that could be associated to anomalies in ovarian function or to malignancy (ovarian, adrenal, pituitary);
- An increase in free estrogen circulating levels, due to reduction of sex binding globulin;
- Alterations in hormones hepatic catabolism;
- An increase in breast hormonal receptors sensibility or number, with normal hormones serum level.

Hyperprolactinaemia has been associated with mammary hypertrophy, but its exact role is not absolutely clear. Not all patients with hyperprolactinaemia present with breast hypertrophy, but patients with pregnancy-induced gigantomastia usually respond well to high dose of an anti-prolactin agent such as bromocriptine. In contrast, breath enlargement may not be arrested in patients with juvenile hypertrophy and hyperprolactinaemia.

Another reported association is between hypercalcemia and juvenile and pregnancy-induced hypertrophy, which has been attributed to an excessive production of PTHrP (parathyroid hormone-related protein). The reason for the increase in PTHrP production that accompany pregnancy-induced hypertrophy is yet not known.

In favor of the hypothesis of an altered hepatic metabolism, one case of macromastia in an infant with Alagille's syndrome has been reported in literature, in which mammary hypertrophy was directly correlated to alterations in the hepatic metabolism of estrogens, rather than to their hyperincretion under a gonadotropic stimulus, to the aromatization of androgens or to central nervous system abnormalities.

Only a few studies exist on juvenile hypertrophy; while much more data are available about pregnancy-induced mammary hypertrophy.

To date any study has confirmed the more accredited hypothesis, that of an increased responsiveness of breast tissue to circulating hormones (receptor hypersensibility).

In literature, several cases of mammary hypertrophy have been reported as adverse effect of a pharmacological treatment. Some authors reported the onset of gigantomastia after prolonged therapy with indinavir, a protease inibitor, used in the treatment of HIV infection.

A Hiv-positive woman, one mounth after beginning a triple therapy with lamivudine, stavudine and indinavir, presented a rapid and progressive increase in breast volume, that regressed after discontinuation of indinavir alone.

The importance of the role of indinavir for this particular case of gigantomastia is confirmed by several considerations:

- Serum level of estrogens or prolactin are within the normal range;
- Indinavir frequently causes non specific morphological changes of some body segments (e.g., abdominal globosity);
- Discontinuation of indinavir alone, while receiving the other drugs, leads to a complete remission of the clinical picture.

Another rare condition is gigantomastia in patients with rheumatoid arthritis, receiving D-penicillamine. The first case has been described by Desai in 1973.

It is likely that, reducing circulating levels of sex hormon binding globulin, D-penicillamine, induces an increase in serum levels of estrogens, thereby determining mammary hypertrophy, or that, chelating zinc ions, sensitizes breast tissue to the action of prolactin or other hormones. A direct action on the mammary gland is also to be considered. Discontinuation of D-penicillamine leads to an arrest in breast growing.

A rare cause of mammary hypertrophy is Systemic Lupus Erythematosus (SLE). In 1960 Shelley described a case of severe bilateral mammary hypertrophy in a young woman with positive SLE tests, diffuse annular erythema and melanodermia.

Propper reported the case of a woman with SLE with disease flare during pregnancy and breast hypertrophy complicated by severe necrotic and ulcerated skin lesions, probably attributable to cutaneous vasculitis. This condition resolved almost completely after delivery under steroid therapy.

Both juvenile and pregnancy-induced mammary hypertrophy related to SLE have unclear etiologies; it is likely that SLE may induce the production of substances that directly or indirectly, by mimicking the action of estrogen or other growth factors, determine an increase in breast volume.

Obesity plays an important role in breast hypertrophy pathogenesis; breast volume increases in all overweight conditions, and it is not surprising that obese patients represent 2/3 of macromastia cases.

In literature it is reported that hypertrophic breast is composed primarily of adipose and fibrous tissue, while the glandular component remains essentially stable.

Lejour reported a mean of 48 percent fat by weight in breast reduction specimens. She also observed that the body mass index has more influence than age on the amount of breast fat.

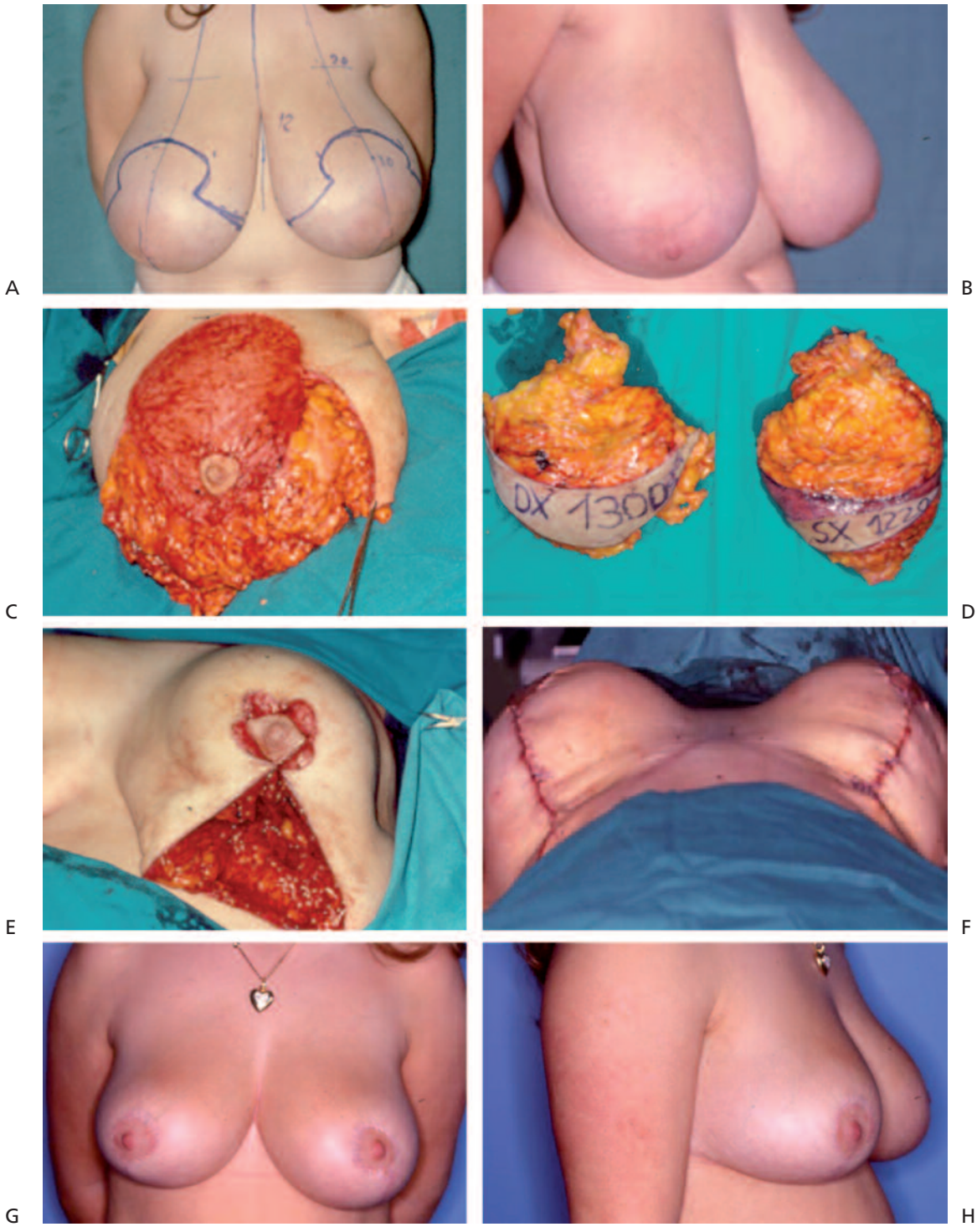

Fig. 19-2. Authors' technique. *A,* Skin markings. *B,* Preoperative view of the patient. *C,* The superior pedicle. *D,* Wedge resection of the inferior pole. *E,* Suture of the lateral pillars. *F,* Immediate postoperative view. *G-H,* One-year postoperative view of the patient.

This pathologic finding has also been highlighted by Strömbeck, who proposed the term "macromastia" for patients requiring reduction mammaplasty, considering incorrect a diagnosis of "mammary hypertrophy".

Several authors confirmed the importance of the fat component in the enlarged breast, and reported their experience with liposuction as an integral part of surgical treatment of macromastia.

TREATMENT

Reduction mammaplasty is the best therapeutic approach for patients with mammary hypertrophy, for both physical and psychological reasons. Its goal is to achieve a reduction of breast volume, while maintaining vascularization and innervation of the nipple-areola complex, and limiting scars.

The first attempts at reducing breast volume were purely functional, and it is only in the last century that aesthetic considerations were taken into account.

Up to 1960, techniques of breast reduction were not safe, because of extensive skin and glandular undermiming. In 1957, Arie realized that it was preferable to avoid any undermiming between skin and gland. In 1963, Skoog proposed an inferoposterior resection and the transposition of the nipple-areola complex on a laterally based dermal pedicle flap, laying the basis for the development of modern reduction mammaplasty.

Since then, many surgical techniques have been got ready, with an increasing interest in reducing scars' lenght.

The choice of the technique depends on the size of the breast, the degree of ptosis, the patient's goal, and surgeon's preferences. The two main decisions that confront the surgeon are the choice of incision pattern, and the choice of pedicle type, which are, for the most part, independent variables.

Different pedicle nipple-areola flap can be employed, including a superior pedicle, an inferior pedicle, a vertical bipedicle, a central mound pedicle, a lateral and a superomedial pedicle.

The incision pattern usually consist of a vertical or an inverted-T scar, which can be applied to any pedicle type, with the advantage of reducing scars' length.

The choice of the pedicle usually depends on the need for elevation of the nipple-areola complex, and the desire to preserve sensory innervation or lactation.

AUTHORS' TECHNIQUE

(Figure 19-2)

An accurate marking of the skin is made preoperatively, which basically refers to Lejour's vertical scar technique; for this purpose the jugular notch is marked, together with two points on the clavicle 5-6 cm at each side of it. A line passing through the nipple is drawn from these points to the inframammary fold (breast meridian).

The new nipple-areola complex position is drawn at the intersection of the inframammary fold and the breast meridian, that usually correspond to a distance of 19-22 cm from the jugular notch, varying with patient height. The areola region is marked with circular patterns on photographic film, obtaining a periareolar circumference of between 14 and 18 cm. In case of gigantomastia an elliptical pattern of 18 cm is used.

The lateral margins of the area to be de-epithelialized are marked by turning the breast in a clockwise direction and counterclockwise, respectively. The lower edge of the marking is obtained by combining the two lateral branches with a circular arc passing 2-4 cm above the fold.

The lower breast quadrants are then infiltrated with saline and adrenaline 1:100,000. Next, de-epithelitation of the upper portion of the marked periareolar area is performed, and extended about 4 cm inferiorly in order to maintain the arterial and venous periareolar network; the skin is dissected medially and laterally as necessary, and the dissection is carried down to the fascial plane.

Once identified the fascial pre-pectoral plane, breast is separated from the fascia of the pectoral muscle along a wide central tunnel of about 8 cm, resection of the inferior pole and of a wedge below the de-epithelialized area is performed.

The pedicle nipple-areola flap is then anchored high to the chest and the two remaining medial and lateral pillars are sutured to each other in order to reassemble the cone breast. At this point, excess skin is evaluated and the direction of the scar is decided.

The remaining skin is defatted very carefully.

We found that in young patients with good skin retraction capability and for resection of up to 1 kg a simple vertical scar can be used.

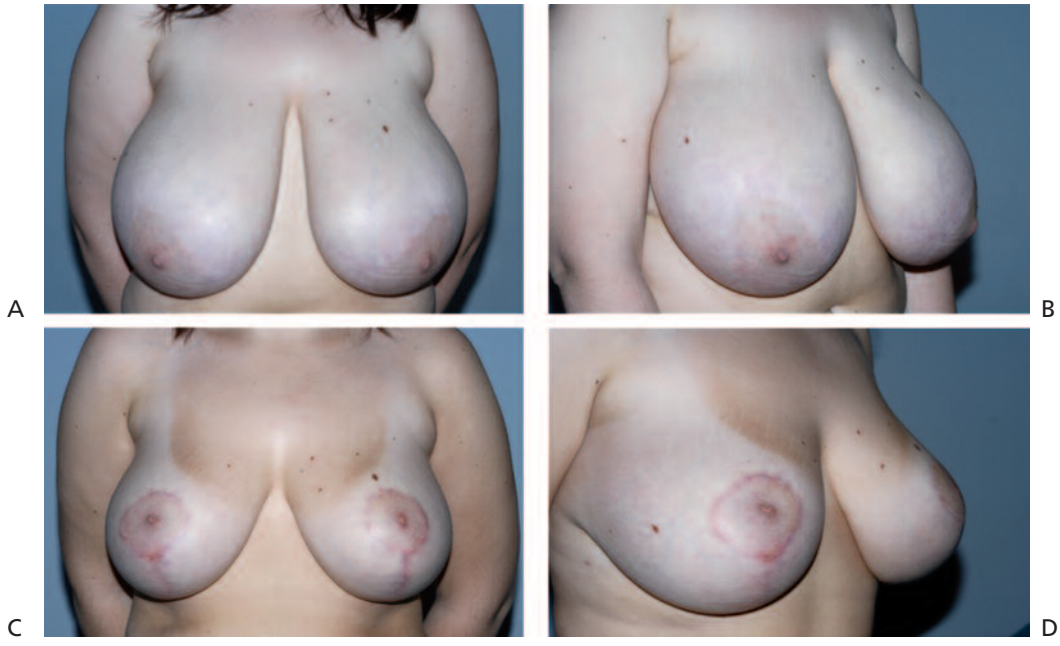

Fig. 19-3. A 25-year-old patient. *A-B,* Preoperative view. *C-D,* One-year postoperative view (Authors' technique).

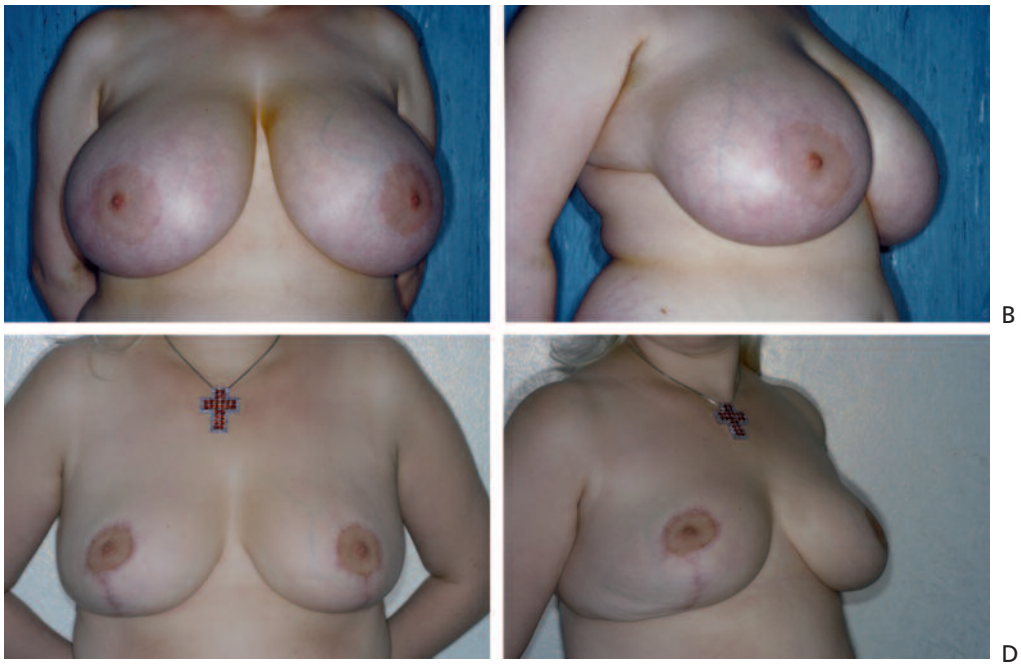

Fig. 19-4. A 28-year-old patient. *A-B,* Preoperative view. *C-D,* Six months postoperative view (Authors' technique).

Rippled appearance of the scar is resolved within a few weeks. In patients with flaccid skin and for resections of over 1 kg with elevation of the nipple-areola complex of over 10 cm, sometimes it is aesthetically more convenient to make a L or T-inverted scar.

The final direction of the scars and the quantity of skin to be removed are decided on a case by case basis (Figures 19-3 and 19-4).

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