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Development of the mammary glands begins in the male and female embryo in identical fashion. During the fourth to fifth week of fetal development, primitive milk streaks, also known as galactic bands, form [1–4]. These are single, thickened ridges of ectoderm that extend bilaterally from the axillary to the inguinal region. Each band consolidates to form a mammary ridge on the thorax, and the remaining band regresses. At 6–8 weeks, a primary bud forms, with thickening of the mammary anlage, which penetrates into the chest wall mesenchyme. The primary mammary bud gives rise to secondary buds that extend into the surrounding connective tissue and become the lactiferous ducts and their branches. The mesenchyme surrounding the duct systems becomes the fibrous stroma and fat of the breast. Between 12 and 16 weeks of development, mesenchymal cells differentiate into the smooth muscle of the nipple-areolar complex (NAC), and branches link to future secretory alveoli. The secondary mammary anlage then develops with differentiation of hair follicles and sweat glands. Between 20 and 30 weeks, placental sex hormones induce canalization of the branched epithelial tissues. By 32–40 weeks, the parenchyma differentiates into alveolar and lobular structures. The epidermis at the origin of the mammary gland becomes depressed, forming a shallow mammary pit onto which the lactiferous ducts open, becoming the NAC. The breast bud becomes palpable at 34 weeks, measuring approximately 3 mm at 36 weeks of age and 4–10 mm by 40 weeks (Fig. 2.1). Soon after birth, the nipple rises because of proliferation of the mesenchyme underneath, and the areola becomes pigmented. Under the influence of maternal hormones that pass into the placenta, male and female neonates may secrete colostral milk, also known as witch’s milk, up to 4–7 days postpartum. Neonates also may demonstrate hyperplasia of the breast, which typically regresses within a few weeks or months of life.

Intrauterine development progresses autonomously and is governed by epithelial-mesenchymal signaling, unlike development in puberty and pregnancy, which depends primarily on hormonal stimulation [4]. Various growth factors regulate mesenchymal-epithelial interactions to guide development [5, 6]. There is good evidence to support that formation of the mammary mesenchyme is directed by signals from the epithelial bud. Transforming growth factor alpha (TGF-α) stimulates ductal and lobulo-alveolar development. TGF-β affects canalization of ductal structures and suppression of lactation. Inhibin and activin are members of the TGF-β family that lead to mammary duct elongation and alveolar development [6]. IGF-1 impacts ductal growth and is expressed in mammary stroma [6, 7]. Laminin-5 aids in hemidesmosome attachment and signaling. Hepatocyte growth factor/scatter factor enhances ductal end bud size, numbers and branching. It is mitogenic for luminal cells and morphogenic to myoepithelial cells [6, 8]. Estrogen is critical for epithelial cell proliferation and ductal morphogenesis [9]. The presence of matrix metalloproteinase (MMP) and the absence of tissue inhibitor of metalloproteinases (TIMP’s) allow necessary disruption of basement membrane and the involution process after weaning. BCL-2 and parathyroid hormone-related protein are other factors that signal the growth and development of the mammary gland [10].
From birth until puberty, the breast remains largely unchanged. Breasts are identical in boys and girls until puberty. Breast development (thelarche) in girls is usually the first sign of sexual maturation. The mammary glands in males normally undergo no postnatal development. Puberty typically occurs at 9 to 13 years of age [11]. At puberty, the breasts rapidly grow and mature under the influence of elevated estrogen, progesterone, and prolactin levels and growth hormones, including luteinizing and follicular-stimulating hormone (LH and FSH), which stimulate estrogen secretion as well as hypothalamic gonadotropin-releasing hormone [6]. Elevated estrogen levels stimulate ductal growth and branching, whereas progesterone influences lobular and alveolar development. Testosterone and dihydrotestosterone, which are androgens, limit breast development [12]. Prolactin stimulates the alveolar buds. Thyroxine also plays a regulatory role. The volume and elasticity of the connective tissues increase, as does the vascularity and fat deposition. Progressive enlargement of the breasts occurs. Other signs of puberty typically follow the onset of thelarche.

Three major periods of the breast life cycle occur after puberty. The first is breast development from adolescence until approximately 25 years. Both stromal and lobular units develop during this period. Pregnancy increases breast weight, with involution postpartum. The female breast further remodels after lactation. After age 35 years, involution occurs with fat replacing breast tissue [9]. With significant decreases in estrogen levels at menopause, ninety percent of the epithelium undergoes apoptosis and fat cells replace breast tissue [12, 13].

Fig. 2.1 Embryonic development of the mammary glands. (a) Ventral view of a 28-day embryo, with regression of the mammary ridge by 6 weeks, as represented in (b). (c-f) Cross sections of the developing breast bud from 6 weeks to birth. (From [2], with permission)
2.1 Anatomy of the Breast

The breasts are situated superficial to the pectoralis major muscle and are hemispheric in shape with an elliptical base in the average young woman. Although breasts vary markedly in size, they normally extend between the second and sixth ribs, vertically, and horizontally between the lateral edge of the sternum and the midaxillary line (Fig. 2.2).

The three major components of the breast include skin, subcutaneous tissue and breast tissue, including parenchyma and supporting stroma. The breast gland is firmly adherent to the skin by suspensory ligaments of Cooper. These fibrous bands, which traverse and support the breast, connect the skin and the deep fascia overlying the pectoralis major muscle.

Lactiferous ducts open on the nipple, and each drains a lobe. The lobes are arranged radially around the breast. Each lobe consists of 20–40 lobules, separated by connective tissue and fat. Each lobule contains 10–100 alveoli. Under the areola, each lactiferous duct has a dilated portion called the lactiferous sinus in which milk accumulates during lactation.

The arterial supply of the breast includes the internal mammary and lateral thoracic arteries, as well as lateral and anterior cutaneous branches of the intercostal arteries from interspaces three, four, and five, and subdermal vessels. Venous drainage flows primarily into the axilla, with further drainage into the internal thoracic, lateral thoracic and intercostal veins. Most of the lymph drains into superficial and axillary nodes. The second to sixth intercostal nerves, chiefly the fourth lateral intercostal nerve, innervate the breast gland and overlying skin.

2.2 Premature Thelarche

Premature thelarche is a benign condition describing premature breast development before the age of 8 years. Premature thelarche is especially prevalent during the first 2 years of life and often resolves during childhood. This condition is typically isolated and rarely progresses to precocious puberty, which is maturation of the hypothalamic-pituitary-gonadal axis with development of two or more sexual characteristics [12, 14]. Volta et al studied 119 girls with premature thelarche, and found that 80% presented prior to 2 years of age, and 60% regressed completely [15]. Some studies report differences of the hormonal milieu of girls affected by premature thelarche. Bioassays have found higher levels of estrogen in girls with premature thelarche. An activating mutation in the GNAS gene, which codifies for the alpha subunit of G stimulating protein, has also been reported [14, 16]. Increased FSH-driven follicular development and mutations in the FSH receptor with higher than normal response levels have also been hypothesized [17–19]. These finding suggest premature thelarche may be an incomplete form of precocious puberty. Phenotypically, girls may also demonstrate accelerated growth and bone age, but are otherwise medically and sexually normal.

Premature thelarche may result from gonadotropin-dependent or gonadotropin-independent estrogen formation, as well as increased sensitivity of estrogen receptors and increased aromatization of adrenal precursors. Exogenous estrogen exposure such as that from cosmetics and hair products, and hormones used in stockbreeding, may serve as an endocrine disruptor [17–19]. Serum levels and metabolic clearance rates of estrogen may be low in children leading to significant effects with exposure that might not be as significant for adults [20]. The possibility of endocrine disorders resulting from hypothalamic lesions, ovarian granulosa cell tumors, follicular cysts, adrenocortical tumors, syndromic and medicinal etiologies must be excluded, and demand a thorough history and physical examination [21].

Premature thelarche must be distinguished from precocious puberty. Characteristics of precocious puberty include estrogenization of vaginal mucosa and labia minora, body odor, pubic and axillary hair, acceleration of growth and rapid bone maturation [12]. No more than 18% of girls with premature thelarche go on to develop precocious puberty [19]. If precocious puberty is suspected, the child should be monitored clinically and referred to a pediatric endocrinologist.

2.3 Breast Masses

Before puberty, it is not unusual to have nodular growth of one or both breasts in either sex. Up to 90% of neonates of both sexes may have palpable breast tissue that may increase in size after birth, but this typically resolves within the first few months after birth [12]. Nodules are typically soft, mobile, and uniform, and
Fig. 2.2 Anatomy of the female breast. (From Netter, FH. Atlas of human anatomy. Summit, NJ: CIBA-GEIGY Corporation, 1989: Plate 167, with permission)
they tend to disappear spontaneously after a few weeks or months; so observation is recommended. Tumors of the pediatric breast are generally benign and rarely may be malignant [22].

Possible causes of breast masses in children include fibroadenoma, hemangioma, lymphangioma, lipoma, abscess and fat necrosis after trauma [23, 24]. Fibroadenoma is the most common breast tumor in pubertal females [12]. Four types of fibroadenomas exist: common fibroadenoma, giant fibroadenoma, juvenile fibroadenoma and phylloides tumors. Common fibroadenomas are most prevalent and present between 14 and 16 years of age. Fibroadenomas usually have a firm, rubbery feel on examination, are mobile, non-tender, and have well-demarcated borders. Cystosarcoma phylloides tumors, which only represent 0.4% of all adolescent breast masses, may reach 20 cm in size [25].

Biopsy of the prepubertal breast may irreversibly hinder later development and is rarely required for diagnosis [24]. Careful physical examination is recommended. Imaging studies like ultrasound may follow, and if there is concern, fine needle aspirate is suggested. Excisional biopsy should be performed in cases of persistently painful or rapidly enlarging lesions, and in children who have a history of malignancy [24, 26] (Fig. 2.3).

### 2.3.1 Gynecomastia

Gynecomastia is the most common form of breast hyperplasia, appearing in 30–57% of healthy men [27]. The term gynecomastia stems from the Greek words gyné (woman) and mastos (breast), and describes female-like enlargement of the male breast leading to glandular proliferation [12] (Fig. 2.4). Gynecomastia presentation may be unilateral or bilateral, and may or may not be associated with pain.

Gynecomastia occurs at 3 time intervals: the neonatal period, adolescence, and old age. Up to 60% of males may develop gynecomastia during adolescence, with peak incidence at mid-puberty. About 75% resolve within 2 years of onset with the rest persisting into adulthood [12, 28].

Gynecomastia may be classified according to the amount of glandular tissue, such as glandular, true gynecomastia; fatty glandular; and simple fatty...
gynecomastia, known as pseudogynecomastia. With true gynecomastia, a firm, rubbery mass may be palpated just below the NAC. The Simon classification of gynecomastia is as follows: Grade 1 is characterized by moderate breast enlargement without skin redundancy; Grad 2a by moderate breast enlargement without skin redundancy; 2b by moderate breast enlargement with marked skin redundancy; and Grade 3 with both marked breast enlargement and skin redundancy [29].

Gynecomastia is most often physiologic, but may manifest a pathologic condition. Gynecomastia is thought to result from serum imbalance in, or production of, estrogens and androgens [27]. There may be a lag in testosterone secretion leading to greater estrogen effect [12]. In adolescents, pediatricians and/or endocrinologists should assess the possibility of hormonal etiology of gynecomastia to reveal possible pathologic conditions, such as hyperthyroidism, congenital adrenal hyperplasia, testicular tumors and hypogonadotropic hypogonadism [12]. A unilateral, firm, fixed mass with overlying skin changes is suspicious for cancer, and should be explored as a possible etiology through mammography or diagnostic fine needle biopsy.

Medical and surgical treatments are available for gynecomastia. Indications for treatment include psychosocial stress and pain, as well as concern for malignancy. While medical treatment is less invasive, it is often ineffective. The basis of medical treatment is hormonal manipulation, which may result in undesirable side-effects. Testosterone and danazol increase androgen level; clomiphene citrate and tamoxifen are antiestrogens, and testolactone is an aromatase inhibitor [30, 31]. Surgical treatments provided by plastic surgeons include liposuction, breast tissue resection and skin reduction [32–35].

2.3.2 Accessory Breast Tissue: Polymastia/Polythelia

Occurrence of accessory breast tissue is most often sporadic, but is familial in 10% of the affected population. Occurrence averages between 0.22 and 6% of the general population. Women have a higher rate than men [36].

The most common type of accessory breast tissue is polythelia. Polythelia, the presence of supernumerary nipples or nipple areolar complexes, is the most common anomaly of the pediatric breast and is found in both boys and girls (Fig. 2.5). Polythelia may occur at any point along the embryonic milk line, from axilla to groin. The condition is both sporadic and familial. Sporadic cases may be associated with nephrourologic abnormalities, and polythelia should therefore heighten suspicion of possible renal abnormalities [22, 36]. Cardiovascular problems associated with polythelia include high blood pressure and conductive or rhythm disturbances [37]. Surgery is requested for esthetic reasons or due to discomfort.

Polymastia is the presence of supernumerary breasts. When a mass is located along the milk line from axilla to groin, the possibility of breast tissue should be considered. Aberrant breast tissue may be found off this axis including the face, neck, torso, vulva, and lower extremities [36]. A common site of ectopic breast tissue is the axilla [37] (Fig. 2.6). Resection should occur prior to puberty to avoid possible glandular development, with elliptical excision sufficing for surgical treatment [22]. The accessory breast tissue often

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**Fig. 2.5** Adolescent male presenting with bilateral polythelia
Congenital and Developmental Abnormalities of the Breast

manifests itself symptomatically during menstrual periods or pregnancy when the breast tissue becomes tender, enlarged, or lactates. Fine needle aspiration confirms diagnosis \[38\]. Surgery may be performed when the breast mass causes discomfort due to tenderness or when secreting milk \[37\].

2.4 Congenital Breast Hypoplasia/ Aplasia

2.4.1 Poland Syndrome

Poland Syndrome is the most frequent cause of congenital breast aplasia or hypoplasia. It presents as a spectrum of congenital deformities of the chest wall, breast and upper extremity in a unilateral fashion (Fig. 2.7). Defined by unilateral absence of the sternocostal head of the pectoralis major muscle, this syndrome was named by Clarkson for Alfred Poland who published his findings in 1841 \[39, 40\]. The syndrome occurs sporadically in 1 of every 20,000–30,000 live births \[22, 41\]. Men are affected more frequently than women (3:1), and the right side is more often affected than the left (3:1) \[42\]. No cases of bilateral involvement have been reported. Renal hypoplasia, certain leukemias and Mobius syndrome have also been associated with the chest wall defects \[43\]. A great spectrum of clinical presentations exist, ranging from mild, with hypomastia and pectoral hypoplasia, to severe, with lack of pectoral major and minor muscles, high insertion of rectus abdominis muscle, paucity of subcutaneous tissue, alopecia of the axilla, rib deformities

Fig. 2.6 Young woman with polymastia with accessory axillary breast tissue

Fig. 2.7 (a) Poland’s deformity with aplastic breast and absent anterior axillary fold as a result of absent pectoralis major muscle. (b) A latissimus musculocutaneous flap was performed to reconstruct the involved breast. This woman will need further tissue to better approximate her contralateral breast.
(II to IV or III to V), sternal rotation, amastia, superior disposition of the NAC and anomalies of the upper extremity including brachysyndactyly [41, 42, 44].

Most cases are sporadic and not familial. It is hypothesized that the etiology of Poland Syndrome is related to an intrauterine accident. One theory is vascular: that the subclavian blood supply is interrupted during limb bud development in the sixth week of gestation, known as subclavian artery supply disruption sequence, disrupting normal development of the chest wall and upper limb [41, 45, 46]. Another theory is related to abnormal migration of embryonic tissues. In a 9-mm embryo, the limb bud that forms the pectoralis muscle develops; by the time it becomes a 15-mm embryo, the bud splits into clavicular, pectoral and sternal components. Faulty attachment or failure of attachment of this primitive limb bud to the upper rib cage and sternum would explain Poland deformity [44, 47].

Patients may desire reconstruction by a plastic surgeon to improve abnormal contours of the chest, anterior axillary fold and breast. Whereas men desire treatment for asymmetry and lack of soft tissue fill on the upper chest, women desire the provision of a symmetrical breast mound, a natural appearing NAC, infraclavicular fullness and a normal anterior axillary fold [43, 46]. While implants have been the mainstay of breast reconstruction in Poland syndrome [48], autologous options include latissimus myocutaneous flap reconstruction possibly with an expander or implant [49], microvascular free flaps including perforator flaps [46, 50] and fat grafting [51]. Latissimus muscle has been traditionally used to simulate the pectoralis major head and anterior axillary fold, and to fill the upper chest, while also providing skin [49, 52]. Endoscopic techniques with minimal scar may be applied to implant placement or latissimus flap transposition [41]. Autologous rib grafting, costal cartilage resection and/or sternal osteotomy may be required [43, 44]. Contralateral, symmetry procedures such as mastopexy might also be necessary.

2.4.2 Tubular/Tuberous Breast

Tubular breast is a term first coined by Rees and Aston [53]. The breast has normal function but abnormal morphology. Tuberosus breast deformity describes a hypoplastic breast with constricting ring around the base of the breast, breast tissue herniation into the areola, deficient skin envelope and inframammary fold malposition (Fig. 2.8). With the narrowed transverse breast diameter and base constriction, the breast appears to herniate into an oversized and protuberant areola [53, 54]. As a result of the breast’s appearance, another name for tubular breast deformity is the Snoopy-nose deformity [55]. The condition may be unilateral or bilateral, and exact incidence is unknown.

Patients consult with plastic surgeons to correct their deformity. Treatment objectives include expanding the base circumference and the skin of the lower hemisphere, releasing constricting skin tightness at the areolar junction, lowering the inframammary fold, increasing breast volume and height and decreasing areolar diameter [54, 56]. A periareolar approach

Fig. 2.8 Tubular breast deformity with narrowed base diameter and pseudo-herniated breast tissue through an enlarged nipple-areolar complex (NAC)
allows alteration of the areolar diameter and division and widening of breast tissue to increase breast base diameter. A tissue expander or implant under the divided breast tissue assists in improving deficient breast volume [57–60].

### 2.4.3 Idiopathic Asymmetry

The initiation of thelarche may occur on one side and proceed at a faster rate for unknown reasons. In most cases, both breasts become relatively equal in volume by the end of puberty. A small degree of breast asymmetry is not uncommon or abnormal; however, a marked inequality of breast volume can be noticeable (Fig. 2.9). Hueston noted that patients experience difficulty in concealing asymmetry greater than 33%, with everyday attire [61].

Idiopathic breast asymmetry is classifiable into six categories: unilateral hypoplasia, asymmetrical hypoplasia, unilateral hyperplasia, asymmetric hyperplasia, hyperplasia/hypoplasia and hypoplasia associated with chest wall deformities. Unilateral hypoplasia is most common and may vary from the minimal idiopathic form to severe Poland syndrome. Associated with breast hypoplasia is a small and cephalad-displaced NAC, and in rare instances, the NAC is absent. Etiologies of breast asymmetry have been described, including differential end organ response to hormonal stimulation during development, tumors, medications and iatrogenic causes, including operations, radiation and trauma [62, 63].

Breast asymmetry may cause physical discomfort as well as psychiatric embarrassment. Early surgical correction may be warranted. Postponing corrective surgery for adolescents with significant asymmetry may be psychologically detrimental and unnecessary. Plastic surgeons use reconstructive techniques to create improved symmetry. Hypoplastic breasts are augmented and may require tissue expansion as a first stage. Tissue expansion preceding placement of a permanent implant allows descent and expansion of the hypoplastic breast and NAC, as well as expansion of the deficient soft-tissue envelope [62]. Particularly in young patients, the expansion process may take place over years until the opposite, unaffected breast reaches maturity [62]. In more severe cases of hypoplasia, the latissimus may be transposed over top of the implant to improve contour and decrease risk of contracture. Correction or camouflage of underlying chest wall deformities may be necessary. The unaffected breast may have ptosis requiring a mastopexy to achieve improved symmetry.

### 2.4.4 Amastia/Athelia

Total absence of the breast is called amastia, and absence of the nipple is athelia. Amastia and athelia result when the mammary ridges fail to develop or completely disappear [25]. The first recorded reference to amastia was in “The Song of Solomon” in the Bible: “We have a little sister, and she hath no breast: What

![Fig. 2.9](a) Idiopathic breast asymmetry with left-sided ptosis and contralateral hypomastia. (b) Postoperative result with left mastopexy and right breast augmentation
shall we do for our sister in the day that she shall be spoken for [1]?” The fictional Amazonian nation was comprised of independent women who removed one of their breasts to gain a competitive advantage in archery. In 1939, Froriep first reported a case of amastia [64]. Amastia has been reported as an isolated finding and a syndromic component. Trier reviewed the literature extensively in 1965 and noted three presentations after reviewing 43 patients: bilateral amastia with congenital ectodermal defects, unilateral amastia and bilateral amastia with variable associated anomalies. Associated abnormalities include cleft palate, hypertelorism, anomalous pectoral muscles, upper limb deformities and abnormalities of the genitourinary tract [64]. Amastia is often associated with anomalous chest wall development, such as in Poland Syndrome (Fig. 2.7). Syndromes with amastia include ectodermal dysplasia, an autosomal dominant hereditary disease, and Mayer-Rokitansky-Kuster-Hauser syndrome with vaginal-uterine agenesis [65–67]. Familial cases have been reported, and inheritance is believed to be autosomal dominant in those cases [1, 64, 68].

Athelia is an extremely rare condition. Absence of the nipples, like amastia, has been reported as a dominant trait in some families and as a finding in various syndromes, including the family of ectodermal dysplasias [69, 70]. Athelia is a component of Al Awadi/Raas-Rothschild syndrome, a lethal autosomal recessive facio-skeletal-genital syndrome [71]. Athelia has also been described as part of the scalp-ear-nipple (SEN) syndrome, an autosomal dominant condition with aplasia cutis congenital, posterior scalp nodules and malformed ears. A case was reported in which the patient had choanal atresia and athelia that was likely induced by methimazole treatment for hyperthyroidism in the pregnant mother [72].

Surgical correction is performed in stages with initial tissue expansion followed by definitive implant. Breast reconstruction may also be performed with autogenous tissue, in particular, free flaps from the abdomen or gluteal region.

2.5 Inverted Nipples

Sir Ashley Cooper first described this entity in 1840. Inverted nipples occur when the tight, shortened deep tissues retract the nipple. Developmentally, this entity originates from a lack of proper elevation of the nipple from proliferation of the mesenchyme underlying the future NAC [73].

Inverted nipples are found in about 2% of the female population and are most frequently bilateral [73, 74]. Although most cases are congenital, acquired causes occur as the result of scarring mastitis, partial mastectomy and prior drainage procedures. Syndromes such as Robinow syndrome and carbohydrate-deficient glycoprotein syndrome include inverted nipples in their constellation of findings [75, 76].

Concerns related to inverted nipples range from esthetic to functional to psychological. Women with this condition may have difficulty breast-feeding. Numerous plastic surgical techniques have been introduced for correction of the inverted nipple [73, 77, 78]. Nipple sensory change, scarring, vascular compromise, obliteration of the ductal system with faulty lactation, and incomplete correction, as well as a high rate of recurrence, may complicate correction.

2.6 Gigantomastia

2.6.1 Juvenile Hypertrophy

Prepubertal hypertrophy is usually bilateral, and virginal hypertrophy developing after puberty may be unilateral or bilateral [22]. Juvenile gigantomastia may be associated with rapid growth to massive proportions in the period surrounding puberty [79]. The breasts are firm and may be nodular. Serum hormone levels are normal, and pregnancy test should be checked to rule this out as a cause. Imaging should be considered to rule out an enlarging mass, and MRI is the preferred modality. Malignancy is rare in prepubertal and pubertal breast at 1.3% [79]. Symptoms associated with significant breast hypertrophy include bra grooving, shoulder, neck and back pain, postural problems, difficulty breathing while supine, and skin necrosis [79]. The physical discomfort coupled with the negative attention and accompanying psychosocial issues result in a very difficult, sensitive situation, which may merit surgical treatment [80, 81].

Goals of surgery are volume reduction with symmetrical breast size and anatomically correct nipple areolar position. Treatment includes breast reduction
techniques. Surgery is best delayed until the end of puberty when breast growth is complete. The risk of recurrence of hypertrophy exists after breast reduction, leading to consideration of hormone therapy or even mastectomy and implant reconstruction [79].

2.6.2 Gravid-Induced Gigantomastia

This entity, though more rare, is similar to virginal hypertrophy, except that rapidly progressive gigantomastia occurs during pregnancy [82–84]. Macromastia may be evident prior to pregnancy, but is exacerbated by pregnancy. Gravid gigantomastia may occur after normal, unaffected pregnancies, but subsequent pregnancies will more likely result in similar gigantomastia. Like virginal hypertrophy, gravid gigantomastia is related to end-organ hypersensitivity to elevated circulating hormone levels, including estrogen and prolactin. A serum factor, like an autoimmune antibody that interferes with the normal hormone-receptor complex, has been proposed [85].

Because of extreme growth, patients experience severe pain, skin ulceration and imminent infection from the wounds. Breasts are tense, firm, and may demonstrate large superficial veins and peau d’orange skin changes. Erosion of veins under excoriated skin threatens hemorrhage.

Either breast reduction or mastectomy is recommended for these patients. Breast surgery performed at the time of pregnancy may threaten viability or developmentally affect the fetus. Some opt for therapeutic abortion, a radical but curative choice [83].

Bromocriptine has been prescribed after delivery to induce involution and, in some cases, during gestation to delay surgical therapy. It lowers secretion of prolactin or may act directly on the breast [85]. Bromocriptine may have teratogenic effects on the fetus [86].

2.6.3 Drug-Induced Gigantomastia

Some cases of gigantomastia are induced by medications. Hormonal therapy, corticosteroids, marijuana, D-penicillamine, cimetidine and the antiepileptic sulpiride may lead to unilateral or bilateral gigantomastia (Fig. 2.10). D-penicillamine, an anti-inflammatory medication prescribed for rheumatologic disorders such as scleroderma, has been frequently reported as a cause for gigantomastia [87–89]. Reversal of gigantomastia occurs with danazol treatment or with stopping D-penicillamine therapy [88]. Medications either stimulate hormones or act locally. Cessation of the potentially offending medication should first be attempted to reverse gigantomastia.

2.7 Deformational Breast Abnormalities: Iatrogenic, Traumatic

Traumas, incisions, infection or radiation to the young female breast may lead to subsequent scarring restricting breast growth. Seatbelt injuries cause compression to skin and underlying fat and may result in breast atrophy and asymmetry [90] (Fig. 2.11). Radiation therapy to the chest wall to treat childhood malignancies ultimately impair breast development [91]. The injured breast nearly always results in hypoplastic deformity, with a combination of deficiencies in skin, nipple areola and/or glandular tissue. Scar tissue at the site of trauma tethers breast tissue to the chest wall, or the injury may result in violation of the breast bud, impeding normal development.

Burn injury may compromise the breast bud or restrict breast growth through constrictive scar...
contracture (Fig. 2.12). Excision and grafting at the initial injury must be performed judiciously to avoid long-term injury and ultimately allow normal growth as burn scar will restrict breast growth and development [92].

Breast and pectoral muscle maldevelopment have been reported in children who have undergone anterolateral and posterolateral surgical incisions though the fourth intercostal space, an approach used for congenital heart surgery [93]. Anterolateral thoracostomies resulted in 60% of the patients subsequently having a greater than 20% discrepancy in volume of the breast and pectoral muscles on the ipsilateral and unaffected sides as reported in a retrospective study from the Children’s Hospital of Pittsburgh. Tube thoracostomy is one of the more common pediatric injuries, and results in scar and fibrous tissue tethering the breast to the chest wall [22].

Great caution must be exercised when creating incisions around the prepubertal breast. Because breast malignancies are so rare in prepubertal patients, biopsy of a suspicious mass is warranted only after a reasonable observation period. Nodular deformities in the breast have been reported after core needle biopsy and diagnosed as reactive spindle cell nodules. These are benign masses resulting from an exuberant reparative response and myofibroblast influx after needle trauma [94].

2.8 Conclusion

Because our culture places importance on breasts and fuels a pervasive fear of breast cancer, individuals with breast and chest wall deformities and breast masses spark strong concerns. Many of these deformities are congenital and based on faulty developmental processes. The breast deformity may be a marker for other
underlying systemic disorders, principally involving the genitourinary and cardiac systems due to parallel development in the embryo. Some breast deformities are iatrogenic, and the potential for damage to the developing breast bud must be considered when considering surgery on the chest of a prepubertal patient.

Although rarely of functional importance, obvious breast deformities may generate devastating psychological stress, causing isolation and withdrawal from social situations. These patients benefit from early consultation with physicians to address their concerns and direct them to treatment to reconstruct their deformity. The reconstructive breast surgeon aims to preserve breast structures and blood supply while achieving improved symmetry and a more normal appearance. Self-esteem is often much improved after surgery even if exact symmetry is not achieved.

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