

Congenital Problems in the Pediatric Breast

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Pediatric Breast

- Embryology
- Post-natal development
- Hyperplasia
- Hypoplasia
- Deformation



Embryology

4th week of gestation: 2 ridges of thickened ectoderm appear on the ventral surface of the embryo between the limb buds



Embryology

By the 6th week ridges disappear except at the level of the 4th intercostal space

Breast Embryology

In other species multiple paired mammary glands develop along the ridges

- Varies greatly among mammalian species
- Related to the number of offspring in each litter



Neonatal Breast

- Unilateral or bilateral breast enlargement seen in up to 70% of neonates
 - Temporary hypertrophy of ductal system
- Circulating maternal hormones
- Spontaneous regression within several weeks



Neonatal Breast

- Secretion of “witches’ milk”
 - Cloudy fluid similar to colostrum
 - Water, fat, and cellular debris
- Massaging breast can exacerbate problem
 - Persistent breast enlargement
 - Mastitis
 - Abscess



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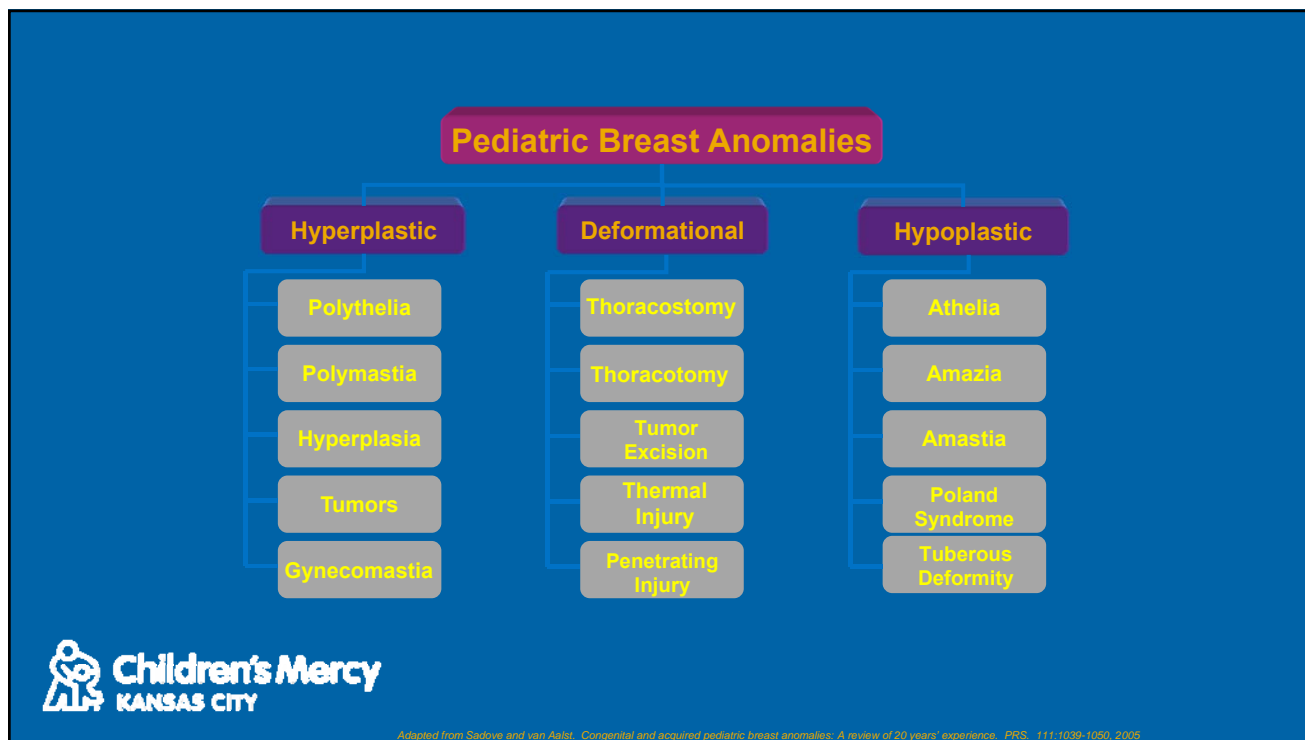
- First stage of normal secondary breast development
 - Average age of 11 years (range 8-15 years)
- Estradiol causes ductal and stromal tissue growth
- Progesterone causes alveolar budding and lobular growth

Pediatric Breast Anomalies

Hyperplastic

Deformational

Hypoplastic



Polythelia

- Supernumerary nipples or nipple-areola complexes
- Most common pediatric breast anomaly
 - Up to 6% of population
- Incomplete involution of the milk line
- Most often in inframammary region

Polythelia

- Males and females
- May be associated with nephrourologic anomalies
- Some familial cases
- Cancerous degeneration possible
- Most commonly inframammary



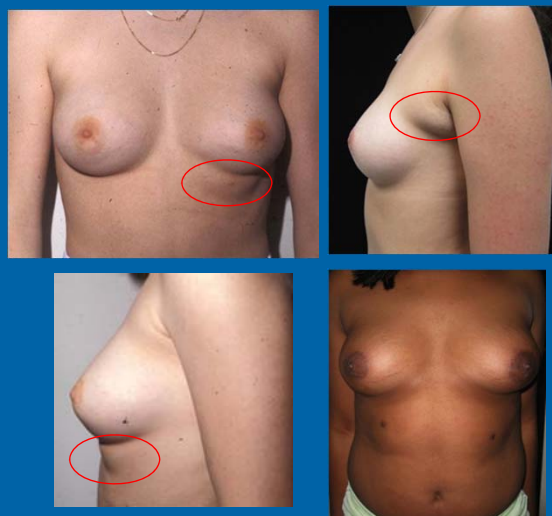
Polythelia

- Multiple nipple-areolar complexes can occur on the breast itself
- MRI may be needed to determine which to preserve



Polymastia

- Accessory glandular tissue
- Less common than polythelia (0.1-1%)
- May occur anywhere along the embryonic milk line
- Often becomes noticeable during puberty, pregnancy, or lactation
- Neoplasia possible



Polymastia

- Treatment requires resection of the accessory glandular tissue
- Follow-up recommended because of possibility of developing cancer in any retained breast tissue



Gynecomastia

- Affects up to 65% of adolescent males
- Peak incidence age 14
- Typically bilateral
- Disk of rubbery tissue beneath nipple
- Tenderness
- Psychosocial distress



Gynecomastia

- Probable transient elevation of estradiol to testosterone ratio
- Ductal and stromal cell proliferation
- Less commonly from hormone secreting tumor, medications, or syndromes



Gynecomastia

- Most begin to resolve in 12-18 months
- Beyond 18 months:
 - Fibrosis and hyalinization occur
 - Less likely to undergo spontaneous resolution
 - More likely surgery will be required



Gynecomastia

Evaluation Includes:

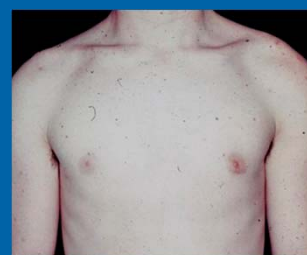
- Complete history
- Rule out endocrine abnormality
- Testicular exam
- Degree of enlargement
- Degree of skin redundancy



Gynecomastia

Treatment Options:

- Reassurance, observation
- Encourage weight loss and exercise
- Drug therapy
- Management of contributing agents
- Surgery
 - Direct excision +/- Liposuction



Macromastia

- Breast development begins with onset of puberty, but reach excessive size
 - Growth disproportional to remainder of the body
- Possible end-organ hypersensitivity to normal gonadal hormone levels
- Wide variability in size
- Can be familial



Macromastia

- Promotes physical symptoms
 - Neck and/or back pain
 - Shoulder grooving
 - Nerve impingement
 - Inframammary intertrigo
- Often severe psychosocial distress
- Limits physical activity >> obesity



Macromastia

- Treatment focuses on
 - Weight loss
 - Physical therapy
 - Surgical breast reduction
- Timing Important
 - Completion of breast growth
 - Increased need for secondary surgery



Weight Associations

- Adolescent breast enlargement can be a deforming, distressing, and disabling condition
- Presenting symptoms and complication rates after breast reduction surgery mirrors those seen in the adult population
- Adolescents may display greater rates of social distress prior to breast surgery



Weight Associations

- The majority of adolescent patients presenting with mammary hyperplasia or gynecomastia requesting surgery are overweight or obese
- More than half of females are obese with a BMI >30 compared to one third of males
- Females are more likely to undergo surgery at a later age than their male counterparts

Weight Associations

- There is a positive correlation between pre-operative BMI and amount of breast tissue removed at surgery
- Post-operatively patients who are followed long-term do not show a significant decrease in BMI with at least three-fourths of both male and female patients remaining overweight or obese



Breast Masses

- Fibroadenoma
- Lipomas
- Hemangiomas
- Hamartomas
- Abscess
- Lymphangioma
- Fat necrosis
- Fibrocystic disease
- Neurofibroma
- Malignancy
 - Primary
 - Metastatic



Breast Masses

- Vast majority of pediatric breast masses are benign
- Most common histology is fibroadenoma
- Observation and follow-up usually reasonable
- Directed work-up for concerning lesions
 - FNA, Ultrasound, mammography, MRI, biopsy



Giant Fibroadenoma

- Benign, discrete lesions
- Present as unilateral rapidly growing breast mass
- Result of localized tissue hypersensitivity to normal gonadal hormones levels



Giant Fibroadenoma

- Needle biopsies may be unrevealing
- Tumors are firm encapsulated masses
- Dense cellular stroma and ductal elements
- Low recurrence risk if adequately resected



Pediatric Breast Malignancy

- Rare: 0.08 per 100,000 in US
- <1% pediatric cancers
- Includes primary, metastatic, and secondary cancers
- All ages, primarily older adolescents aged 15-19
- Variable histologic types:
 - Secretory carcinoma, phyllodes tumors, intraductal carcinoma, metastatic rhabdomyosarcoma, lymphoma, neuroblastoma



Hypoplastic Anomalies

- Athelia – absence of the nipple
- Amazia – absence of the mammary gland
- Amastia – absence of the nipple and gland
- Rare disorders – *Incidence unknown*
 - Unilateral – Poland Syndrome
 - Bilateral - Congenital ectodermal defects
- Familial inheritance
- ? Teratogens



Hypoplastic Anomalies

- Abnormal development or involution of mammary ridge
- Associated with syndrome or multiple anomalies
- Congenital ectodermal defects with abnormalities of skin, appendages, teeth, nails



Tuberous Breast

- Relatively common anomaly
- Males and females
- Unilateral or bilateral
- Poorly understood
- No associated findings
- “Snoopy breast” or “Tubular breast”



Tuberous Breast

- Deficient skin envelope
- Deficient base diameter
- Herniation of breast tissue into areola
- Inframammary fold malposition
- Variable hypoplasia



Poland Syndrome

- Sporadic disorder
- Absence of sternocostal head of pectoralis major muscle
- Brachysyndactyly
- Chest wall hypoplasia
- Breast and nipple hypoplasia
- Loss anterior axillary fold



Poland Syndrome

- Unilateral, rarely bilateral
- Sporadic, rarely familial
- 3:1 Male to female ratio
- Widely variable presentation
- Interruption subclavian artery blood flow 6th week
- Associated renal anomalies



Poland Syndrome

- Incidence 1:20,000-30,000
- Familial cases reported
- Features vary in severity and extent
- 3 : 1 Males : Females
- Right side 60-75%
- Interruption of embryonic blood supply in subclavian artery distribution at 6 weeks



Poland Syndrome

- Absence other chest wall muscles (pectoralis minor, latissimus, serratus, external oblique)
- Deformity/aplasia ribs 2-5
- Deficiency subcutaneous tissues
- Associations with Mobius, Klippel-Feil, renal anomalies, dextrocardia
- Reported cancers including breast cancer on affected side



Poland Syndrome

Surgical Indications:

- Chest wall depression
- Inadequate protection of mediastinum
- Paradoxical movement of chest wall
- Aplasia/hypoplasia of breast
- Cosmetic defects



Poland Syndrome

Treatment Options

- Breast or chest expander/implant
- Muscle or fat flap +/- implant
- Fat grafting
- Nipple repositioning/reconstruction

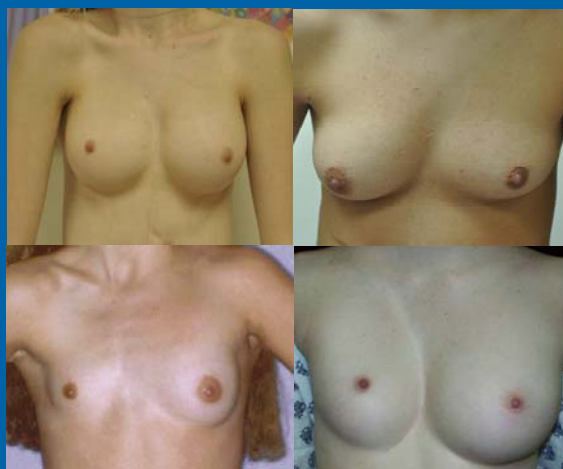
Surgical correction often imperfect

- Residual nipple, contour asymmetries
- High rate of revision with implants



Anterior Thoracic Hypoplasia

- Appears similar to Poland Syndrome
- Chest wall hypoplasia
- Variable breast hypoplasia
- Nipple malposition
- Pectoralis muscles intact
- ? Variant Poland's



Glandular Hypoplasia

- Notable breast asymmetry of varying severity
- Not secondary to any other cause
- More common than Poland syndrome
- Insurance coverage inconsistent



Glandular Hypoplasia

Treatment:

- Breast implant reconstruction
- Adjustable implant with sequential expansion to match growth
- Breast reduction or mastopexy
- Autologous fat grafting



Glandular Hypoplasia

- Milder cases can still significantly impact patient



Iatrogenic Injury to Breast

- Secondary to chest tube placement
- Scar and fibrous tract tethers breast tissue to chest wall
- Requires release to accommodate breast growth



Iatrogenic Injury to Breast

- Secondary to thoracotomy
- Breast tissue tethered to the anterior chest wall
- Violation of the breast bud by the initial incision
- Loss of peripheral muscle bulk



Iatrogenic Injury to Breast

- Secondary to tumor incision/excision
 - Abscess, Cyst, Hemangioma, Lipoma
- Excision prior to breast development may injure breast bud
- Resultant breast hypoplasia
- Resultant tethering



Traumatic Injury to Breast

- Thermal Injury
- Growth hindered by scar contractures
- Loss of anatomy/landmarks
- Initial burn excision and grafting can injure breast bud



Thank You!

