Pediatric Thyroid Surgery

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Objectives

- Discuss surgical congenital thyroid disease
- Identify rare indications for thyroid surgery in benign thyroid disease
- Clarify work-up for pediatric thyroid nodules
- Review treatment for pediatric thyroid cancer

Congenital Hypothyroidism

- Incidence 1:4000
- Overt symptoms may not be present at birth
- Profound effects on brain development
- Work up is:
  - No extensive testing for etiology
  - Does not change neonatal treatment
  - May allow for assessment of future risk for other children
- May not have etiology identified
Ectopic Thyroid

- 1/100,000-300,000
  - Lingual thyroid most common (90%)
  - 70-75% no orthotopic thyroid
- Common sx:
  - dysphagia, dysphonia, globus, cough, snoring/OSA
- Most are hypothyroid
- Rare carcinoma: usually papillary
- Evaluation:
  - MRI, CT, thyroid scan, US, FNA
  - Serial observation

Treatment Approaches: Lingual

- Intervention
  - Concern for malignancy, thyrotoxicosis, obstructive symptoms, dysphagia
- Surgical
  - Transoral: cautery, laser, robotic
  - Transhyoid
  - Suprahyoid
  - Lateral pharyngotomy +/- trach
- Medical
  - Suppression with levothyroxine
  - I-131: higher dose, adults only

Branchial Cleft Anomalies

- Fourth Branchial anomaly
  - implicated in recurrent neck/thyroid abscesses
  - 1-2% of branchial anomalies
  - 97% on the left
  - Cysts or sinuses near or in the thyroid gland
  - Complete fourth fistula never reported
- DDx:
  - 3rd branchial cleft anomaly, thymic cyst, ectopic thyroid, atypical mycobacterium, other congenital
- Studies: Barium esophagram, CT, MRI, US
- Controversy about identification of RLN
- High recurrence without hemithyroidectomy
Goiter

- **Acquired**
  - Colloid
  - Inflammation
  - Iodine deficiency
  - Goitrogen
  - Infiltrative disease
  - Toxic goiter
  - Adenoma
  - Carcinoma

- **Congenital**
  - Thyroglossal duct cyst
  - Branchial cleft anomaly
  - Pendred syndrome
  - Long differential

Hyperthyroidism

- 10-15% of all pediatric thyroid disorders
- 30% of all pediatric goiters
- Graves Disease (>95%) of cases
  - Relatively rare in children
  - Incidence increases with puberty
  - Female: male (3-5:1)
  - Medical treatment is standard
  - I-131 or surgery for antithyroid failure
    - Surgery may be favored due to long-term risk of malignancy, especially in children

Graves Surgical Principles

- Traditional: bilateral near total thyroidectomy
  - Difficult to determine how much remnant to leave.
  - Risk of both hypo- and recurrent hyperthyroidism.
    - Recurrent hyperthyroidism usually treated with I-131.
- New: Total thyroidectomy
  - Must maximize protection of parathyroids.
  - Experience needed to avoid injury to recurrent laryngeal nerves.
  - Hypothyroidism is common sequelae regardless of approach.

Colloid (nontoxic) Goiter

- Diffuse enlargement of thyroid gland
- Typically pubertal years
- Normal thyroid function tests
- Often family history
- May represent mild autoimmune thyroiditis
  - TPO Ab titer may help to distinguish
- Therapy usually not necessary
  - Suppression is controversial
- Surgery possible for obstructive symptoms
Thyroid Nodules

• Low prevalence in children
  – 0.2 – 1.3 % of children (4% of adults)
  – 3rd most common solid tumor in children
  – F:M : higher in young boys and older girls

• Higher incidence of malignancy
  – ~20% (9-50%); 5% in adults
  – The younger the higher
  – Even hot nodules have higher malignancy

• Benign
  – Many same as adult
  – Intra-thyroid thymic tissue

Flexible laryngoscopy

• The nose noodle!

Evaluation

• Labs: TSH, T4, T3
  – Cannot rule out malignancy
  – Antithyroid peroxidase Abs and anti-thyroglobulin Abs may signify increased CA risk
  – Calcitonin \( \rightarrow \) MTC

• Ultrasound can assist in detection
  – Many “single nodules” are
    • Multiple nodules,* cystic, enlarged thyroid lobe, Hashimoto’s with a prominent thyroid, thyroglossal duct cyst, lymph nodes
  – Cannot distinguish benign and malignant
    • Goldbarb, et al., 2012: microcalc, abnormal LN, “tall” nodules \( \rightarrow \) likely to be malignant
  – Uptake scan not reliable

Thyroid Nodules

• Presentation
  – Painless thyroid mass
  – Euthyroid, hyperthyroid or hypothyroid symptoms
  – Painful, overlying erythema, fever

• Concern
  – Airway compression
  – Rapid growth
  – Dysphagia
  – Voice changes
  – Firm, irregular, fixed LNs
  – Rare LN without thyroid mass
FNA

- 3 recent pediatric studies
  - Highly sensitive
  - Recommend use in all ped thyroid nodules
  - Metanalysis:
    - 82% sensitivity, 91% specificity
    - 9-20% non-diagnostic
  - Ultrasound-guided
  - Difficult: nodules <1cm
  - Often need sedation if young

Thyroid Cancer

- Rare
  - 2.7% of all thyroid cancers occur in children
  - Puberty: Most in 15-19 year old range
  - 2:3:1 girls: boys

- Types
  - Papillary carcinoma ~80-95%
  - Medullary carcinoma ~5-15%
  - Follicular and Anaplastic <5%

Thyroid Cancer

- Risk factors
  - Ionizing radiation
    - Thyroid is more susceptible in children
    - Esp < 5 years
  - Iodine deficiency
  - Autoimmune thyroiditis
  - Prolonged TSH elevation
  - FH

Autosomal Dominant Syndromes

- Medullary thyroid carcinoma (MTC)
  - Familial, isolated

- Multiple Endocrine Neoplasia 2a and 2b

- Gardner Syndrome:
  - Thyroid, breast, colon CA, lipomas, intestinal polyposis, osteomas

- Cowden disease:
  - Thyroid and breast CA, multiple hamartomas

- Familial adenomatoid polyposis
Treatment

• Surgery is primary treatment
• Supplemented by
  – I-131, thyroid replacement

Pediatric WDTC

• Substantial controversy on extent of thyroidectomy
  – Especially in children
• Proponents of lobectomy:
  Society of Surgical Oncology
  – Lower complications
  – Does not reduce overall mortality
  – <5% recurrence occurs in the thyroid bed
  – Studies on pediatric recurrence of lobectomy vs. total
    • some with no difference
    • others with recurrence as high as 50%

• Proponents of total/near total thyroidectomy:
  American Thyroid Association, American Association of Clinical Endocrinologists
  – Complications rare in experienced hands
  – Most PTC have micro foci in contralateral lobe
  – Decreases rate of recurrence
  – Radiiodine and thyroglobulin levels for persistent/recurrent disease
  – Completion thyroidectomy has higher rate of complications

• Most centers recommend total or near-total
• Cervical LNs
  – Debated
  – Extensive regional metastases in children at initial presentation (60-80%)
  – Higher distal metastases: 10-20% have lung at dx
Pediatric WDTC

- Follow-up
  - Whole Body Scan/thyroglobulin
    - most often iodine-avid and highly TSH-sensitive
    - 8-12 weeks post-op
    - Life-long, but at longer intervals
  - Survival is 99% at 10 years, 90% at 30 years.
    - Children have higher local and distant recurrence.
    - better survival with extensive disease
    - Progression-free survival:
      - 65-70% at 5 years
      - 46% at 20 years.

Outcomes

- Higher risk
  - <10 years old
  - Distant disease at presentation
  - Positive surgical margins
  - Thyroid capsule invasion, soft-tissue invasion
  - Non-diploid DNA
  - Overexpression of p21 ras or mutations of N-ras

Medullary Carcinoma

- Almost all are MEN II
- Hereditary in 20-30%
- Calcitonin levels aid in diagnosis and follow-up
- MTC is always bilateral and multiple
- Spreads very early
- MEN 2A
  - Not usually clinically apparent before age 12
    - Metastatic MTC seen as early as 3 months of age
- MEN 2B
  - Much earlier MTC
  - Case report of disease at 9 weeks old

MEN

- ret proto-oncogene on chromosome 10
- Neonatal testing in children with FH of MEN2A and 2B
- Early prophylactic thyroidectomy (ATA, 2009)
  - MEN 2A:
    - consider before 5 years of age
    - except codon 634→ must before 5 yo.
  - MEN 2B:
    - ASAP and by one year.
**Surgery**

- Smaller anatomy
- Requires highly experienced surgeons
- Minimally invasive
  - similar to adults in reduced length of stay and incision size

Seybt and Terris, 2011

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**Experience counts**

- Higher volume endocrine surgeons (Sosa, 2008)
  - Significantly shorter stay
  - Lower costs
  - No difference in complications
  - Access to high-volume surgeons is less
    - low-income, Black, or Hispanic

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**Surgery risks**

- < 6 years old are higher risk: 21%
- Hypoparathyroidism (2%)
- RLN injury (1%):
  - 3.8% under 6 years of age.
  - may compensate more easily.
- External superior laryngeal nerve

Sosa, et al., 2008

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**Index case MEN 2B**

- Prenatal genetic counseling
- Prenatal surgical consult, scheduling of anticipated surgery 1 month after due date
- Birth of baby
- RET mutation testing soon after birth
- Positive
- Negative
- Proceed with surgery soon after the first 30 days
- Cancel surgery

Shankar, et al., 2012
Collaborative Approach

- High-volume endocrine surgeon
  - Thyroid = mostly adult surgery
- Pediatric head and neck surgeon
  - Expertise in smaller anatomy of pediatric neck
- Pediatric and Adult endocrinologist
- The University of Wisconsin Endocrine Surgery Center

Wood, et al., 2011

Summary

- Thyroid disease may have wider differential in children.
- Surgery more challenging, especially in very young.
- Pediatric thyroid cancer has excellent outcomes.
- Collaboration is key!

Thank you!

References