# Hyperparathyroidism in Pediatric Age Group

*ESSAY*

**Submitted For Partial Fulfillment of Master Degree In General Surgery**

*BY*

**Mostafa Hussein Ahmed Eltaweel**
**( M.B.B.ch )**

*Supervisors*

*Prof. Dr.* **Osama Abdel Ellah Elnaggar**
**Professor of Pediatric Surgery**
**Faculty of Medicine - Ain Shams University**

*Dr.* **Mohamed Soliman Eldebeiky**
**Assistant Professor of Pediatric surgery**
**Faculty of Medicine- Ain Shams University**

*Dr.* **Amr Abdel Hamid Zaki**
**Lecturer of Pediatric surgery**
**Faculty of Medicine - Ain Shams University**

**Faculty of Medicine**
**Ain Shams University**
**2011**

# فرط نشـاط الغدة الجار درقية في الأطفال

رسالة

**توطئة للحصول على درجة الماجستير في الجراحة العامة**

مقدمة من

الطبيب/ مصطفي حسين أحمد الطويل

بكالوريوس الطب و الجراحة، جامعة الإسكندرية

تحت إشراف

أ .د/ أســامة عبد الإله النجار

أستاذ جراحة الأطفال

كلية الطب – جامعة عين شمس

د/ محمــد ســليمان الدبيكـى

أستاذ مساعد جراحة الأطفال

كلية الطب – جامعة عين شمس

د/ عمــرو عـبد الحمـيد زكـى

مدرس جراحة الأطفال

كلية الطب – جامعة عين شمس

كلية الطب

جامعة عين شمس

٢٠١١

# *Summary*

# SUMMARY

Hyperparathyroidism is the most frequent disorder of the parathyroid in children and may be primary, secondary, or tertiary. Primary Hyperparathyroidism in childhood is uncommon, although many adults with the condition date the origin of their symptoms to late adolescence.

Neonatal Hyperparathyroidism is a rare condition. It is accompanied by high mortality if not identified and treated early. Critical levels of Hypercalcemia may develop within the first several days of life, although occasional infants have a more insidious onset with gradual elevation of calcium levels over several months. The infants often manifest symptoms of lethargy, hypotonia, and dehydration, mild to moderate respiratory distress, slow feeding and failure to thrive.

The surgical management of neonatal hyperparathyroidism was first reported in 1964, and the urgency therefore was recognized shortly after. Initially, subtotal parathyroidectomy was the procedure of choice, but owing to a significant rate of recurrence, total parathyroidectomy was adopted. Total parathyroidectomy was complemented with heterotopic autotransplantation. The success of parathyroid autotransplantation in adult patients has encouraged its use for neonates and children.

In Familial hyperparathyroidism, which is more frequent in children than in adults, diffuse chief cell hyperplasia involves all parathyoid glands. Children with the condition often have associated syndrome complexes including MEN 1, MEN 2A, and familial hypocalciuric hypercalcemia. Most children are recognised during screening of family members at risk for these syndromes. Hyperparathyroidism is often the earliest manifestation of MEN 1.

A few cases of familial hypocalciuric hypercalcemia have been reported that cause a benign form of hypercalcemia, usually diagnosed after age 10 years. This disorder may be transmitted as an autosomal dominant trait. Children with this disorder do not excrete normal amounts of calcium in the urine because of the abnormal renal response to parathyroid hormone. Total parathyroidectomy is usually necessary to achieve normocalcemia, with autotransplantation of one gland in the forearm.

Primary hyperparathyroid disease among pediatric patients is more common in females, is most commonly due to a single adenoma, and is associated with significant morbidity. Despite the high frequency of symptoms in pediatric hyperparathyroid patients compared with their adult counterparts, definitive diagnosis is often significantly delayed after the onset of

symptoms. Children that are suspected of having HPT should be screened by using serum calcium and PTH levels. Additional laboratory and radiographic studies are often helpful for verifying the diagnosis, evaluating for complications of HPT and preparing for surgical treatment. Parathyroidectomy has few complications, is effective at restoring normal serum calcium, and is the treatment of choice in children with HPT.

Despite advances in medical and surgical treatment, the incidence of secondary and tertiary HPT is on the rise because of the increasing incidence and prevalence of CRF. Because of earlier diagnosis of secondary HPT and new medical treatment options, the incidence of parathyroidectomy in this population has been constant or decreasing throughout the world even though the incidence of secondary HPT has increased.

Surgical management of secondary and tertiary HPT is safe and effective at correcting bone mineralization and metabolic disturbances. The most commonly accepted approaches in these patients are subtotal parathyroidectomy or total parathyroidectomy with autotransplantation of parathyroid tissue into the non-dominant forearm. While surgery remains the only cure for patients with tertiary HPT, the treatment of secondary is predominantly medical employing newer calcimimetics, phosphate binders, and vitamin D analogues.

Invasive radioguided parathyroidectomy (IRP) is a very attractive surgical approach to treat patients with HPT. IRP has proven to be technically easy, safe, and with a low morbidity rate in the hands of a skilled surgeon. The advantages of IRP over bilateral neck exploration in patients with HPT can be summarized as follows:

(a) Smaller incision, less surgical trauma;

(b) Shorter length of surgery, anesthesia, and hospital stay;

(c) Less postsurgical pain;

(d) Better cosmetic results; and

(e) Lower overall cost.

# *List of Tables*

# *Contents*

*Chapter* *Page*

# *List of Abbreviations*

| | |
|---|---|
| **BMD** | Bone Mineral Density |
| **Ca/Cr** | Calcium/ Creatinine |
| **CAMP** | Cyclic Adenosine Monophosphate |
| **CASR** | calcium-Sensing Receptor |
| **CRF** | Chronic Renal Failure |
| **CT** | Computed Tomography |
| **cTAL** | Cortical Thick Ascending Limb of the Loop of Henle |
| **DCT** | Distal Convoluted Tubule |
| **EAMIP** | endoscopically Assisted, Minimally Invasive Parathyroidectomy |
| **FDG** | Fluro-Deoxy-Dglucose |
| **FHH** | Familial Hypocalciuric Hypercalcemia |
| **FLE** | Focused Lateral Exploration. |
| **FNAB** | Fine Needle Aspiration Biopsy |
| **FNAC** | Fine Needle Aspiration Cytology |
| **FNE** | focused Neck Exploration |
| **HHM** | humoral Hypercalcemia of Malignancy |
| **HPT** | Hyperparathyroidism. |

| | |
|---|---|
| **IOPTH** | Intraoperative Parathyroid Hormone |
| **MEN** | Multiple Endocrine Neoplasias |
| **MIP** | Minimally Invasive Parathyroidectomy |
| **MIRP** | Minimally Invasive Radioguided Parathyroidectomy |
| **MIVAP** | Minimally Invasive Videoscopically Assisted Parathyroidectomy |
| **MRI** | Magnetic Resonance Imaging |
| **NHPT** | Neonatal Hyperparathyroidism |
| **NIH** | National Institute of Health |
| **NSAID** | Non Steroidal Anti Inflammatory Drugs |
| **NSHPT** | Neonatal Severe Hyperparathyroidism |
| **PET** | Positron Emission Tomography |
| **PHPT** | Primary Hyperparathyroidism |
| **PTH** | Parathyroid Hormone |
| **PTHrp** | Parathyroid Hormone Related Protein |
| **RAI** | Radioactive Iodine |
| **RANK** | The Receptor Activator of Nuclear Factor Kappa. |
| **RANKL** | The Receptor Activator of Nuclear Factor Kappa Ligand. |
| **RGP** | Radio-Guided Parathyroidectomy |

| | |
|---|---|
| **RGVAP** | Radioguided Video Assisted Parathyroidectomy |
| **SCM** | Sternocleidomastoid Muscle. |
| **SPECT** | Single Photon Emission Computed Tomography |
| **U/S** | Ultrasonography |
| **UE** | Unilateral Neck Exploration |
| **VDR** | Vitamin D-Receptors |

# *List of Figures*