HYPOPARATHYROIDISM TREATMENT GUIDELINES

Management of Hypoparathyroidism: Summary Statement and Guidelines were developed by 50 international experts in hypoparathyroidism and general endocrinologists from 15 countries who met over 24 months to review key issues. The guidelines were published in the Journal of Clinical Endocrinology and Metabolism (JCEM) December 2022. It summarizes current information regarding epidemiology, presentation, diagnosis, clinical features, and management of Hypoparathyroidism (HPTH).

Incidence

In the United States there are 37 cases per 100,000 persons. (U.S. population as of 2021 is 331.9 million) That makes approximately 122,000 people with hypoPARA in the U.S.

Permanent hypoPARAthyroidism is said to occur post neck surgery in less than 3% of cases.

Causes

The most common cause (>75%) is postsurgical (following neck surgery).
The second most common cause of HPTH is autoimmune disease of the endocrine glands.
Remaining causes of HPTH include 20+ different genetic disorders that impact PTH secretion or action.

Diagnosis /Presentation

Hypocalcemia in the presence of undetectable or low PTH level
Confirmed on 2 occasions at least 2 weeks apart.
Abnormalities which support the diagnosis:

- Hyperphosphatemia
- Low serum level of 1,25 dihydroxy Vitamin D
- Low bone turnover markers
- History of thyroid or neck surgery
- Pseudo-hypoparathyroidism is associated with resistance to PTH and presents with high PTH and low serum calcium.

Clinical Features

Life threatening, acute symptoms include seizures, laryngospasm, and cardiac arrhythmias.

• Chronic, non-life threatening symptoms include numbness, tingling, muscle cramping, cognitive difficulties (“brain fog”), and a prolonged QT seen on electrocardiogram (EKG).

• Chronic hypocalcemia and hyperphosphatemia (elevated phosphate levels) may lead to increased risk of soft tissue calcifications and kidney stones.
• Bone mineral density is typically higher than average. Low bone turnover is characteristic, causing dense bone that may still be prone to fracture.
• In treated HPTH patients, higher urine calcium levels (hypercalciuria) are seen, due to large doses of calcium and active vitamin D needed.
• Other complications that have been seen include serious infection, cardiovascular disease, cataracts, and fracture of the upper extremities.

G U I D E L I N E S F O R D I A G N O S I S A N D E V A L U A T I O N

- Hypocalcemia (low ionized serum calcium or total serum calcium adjusted for albumin) in the presence of an undetectable or inappropriately low intact PTH (utilizing either a 2nd or 3rd generation assay) on two occasions at least two weeks apart confirms the diagnosis.
- Additional abnormalities caused by low PTH which support the diagnosis: Elevation in serum phosphorus, reductions in 1,25(OH)2D, and elevations in the urinary fractional excretion of calcium.
- In patients with postsurgical HypoPT, panel members regard the condition as permanent if the HypoPT persists > 12 months after surgery.

What is the role of genetic testing in the diagnosis and evaluation of chronic HypoPT?

- In patients with nonsurgical HypoPT who have a positive family history of nonsurgical HypoPT, present with syndromic features, or are younger than 40 years, panel members undertake genetic testing.
- In patients with nonsurgical HypoPT who have other clinical features of autoimmune Poly endocrinopathy–candidiasis–ectodermal dystrophy syndrome (APECED), panel members undertake genetic testing for *autoimmune regulator (AIRE) gene* variants.
- Panel members avoid the designation of “autoimmune HypoPT” for patients who do not have APECED because there are no definitive diagnostic tests for polygenic autoimmune HypoPT.

What are the most common symptoms and complications of chronic HypoPT reported in the literature?

Observational studies comparing patients with HypoPT to controls with normal parathyroid function have identified the following complications associated with HypoPT (percentages represent the median among all studies): cataract (17%), infection (11%), nephrocalcinosis/nephrolithiasis (15%), renal insufficiency (12%), seizures (11%), depression (12%), ischemic heart disease (7%), and arrhythmias (7%)

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