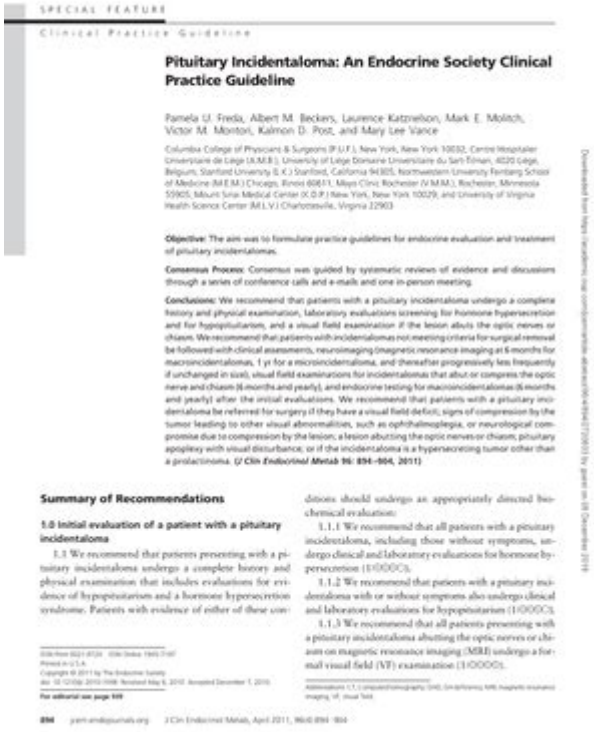
Pheochromocytoma Endocrine Society Guidelines



SPECIAL FEATURE

Clinical Practice Guideline

Pituitary Incidentaloma: An Endocrine Society Clinical Practice Guideline

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Objective: The aim was to formulate practice guidelines for endocrine evaluation and treatment of pituitary incidentalomas.

Consensus Process: Consensus was guided by systematic reviews of evidence and discussions through a series of conference calls and e-mails and one in-person meeting.

Conclusions: We recommend that patients with a pituitary incidentaloma undergo a complete history and physical examination, laboratory evaluations screening for hormone hypersecretion and for hypopituitarism, and a visual field examination if the lesion abuts the optic nerves or chiasm. We recommend that patients with incidentalomas not meeting criteria for surgical removal be followed with clinical assessments, neuroimaging (magnetic resonance imaging at 6 months for macroincidentalomas, 1 yr for a microincidentaloma, and thereafter progressively less frequently if unchanged in size), visual field examinations for incidentalomas that abut or compress the optic nerve and chiasm (6 months and yearly), and endocrine testing for macroincidentalomas (6 months and yearly) after the initial evaluations. We recommend that patients with a pituitary incidentaloma be referred for surgery if they have a visual field deficit, signs of compression by the tumor leading to other visual abnormalities, such as ophthalmoplegia, or neurological compromise due to compression by the lesion; a lesion abutting the optic nerves or chiasm; pituitary apoplexy with visual disturbance; or if the incidentaloma is a hypersecreting tumor other than a prolactinoma. (*J Clin Endocrinol Metab* 96: 894–904, 2011)

Summary of Recommendations

1.0 Initial evaluation of a patient with a pituitary incidentaloma

1.1 We recommend that patients presenting with a pituitary incidentaloma undergo a complete history and physical examination that includes evaluations for evidence of hypopituitarism and a hormone hypersecretion syndrome. Patients with evidence of either of these conditions should undergo an appropriately directed biochemical evaluation:

1.1.1 We recommend that all patients with a pituitary incidentaloma, including those without symptoms, undergo clinical and laboratory evaluations for hormone hypersecretion (1⊕⊕⊕○).

1.1.2 We recommend that patients with a pituitary incidentaloma with or without symptoms also undergo clinical and laboratory evaluations for hypopituitarism (1⊕⊕⊕○).

1.1.3 We recommend that all patients presenting with a pituitary incidentaloma abutting the optic nerves or chiasm on magnetic resonance imaging (MRI) undergo a formal visual field (VF) examination (1⊕⊕⊕○).

ISSN Print 0021-972X ISSN Online 1945-7197
Printed in U.S.A.
Copyright © 2011 by The Endocrine Society
doi: 10.1210/jc.2010-1048 Received May 6, 2010 Accepted December 1, 2010

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pheochromocytoma endocrine society guidelines provide crucial recommendations for the diagnosis, management, and treatment of this rare neuroendocrine tumor. Recognizing the importance of standardized practices, these guidelines serve as an essential resource for healthcare providers working with patients who may be affected by pheochromocytoma. This article will delve into the key aspects of the Endocrine Society guidelines, including the definition and classification of pheochromocytoma, diagnostic approaches, treatment options, and ongoing management strategies.

Understanding Pheochromocytoma

Pheochromocytoma is a rare tumor that arises from chromaffin cells in the adrenal gland, leading to excessive production of catecholamines, such as adrenaline and norepinephrine. This overproduction can result in a range of symptoms, including hypertension, palpitations, sweating, and anxiety. The tumor can be sporadic or associated with genetic syndromes, making it vital for clinicians to follow established guidelines to ensure accurate diagnosis and effective management.

Definition and Classification

According to the Endocrine Society guidelines, pheochromocytomas can be classified based on their genetic and clinical characteristics:

- **Sporadic Pheochromocytoma:** Occurs without any identifiable genetic syndrome.
- **Hereditary Pheochromocytoma:** Associated with specific genetic syndromes, including:
    - Multiple Endocrine Neoplasia type 2 (MEN 2)
    - Von Hippel-Lindau disease
    - Neurofibromatosis type 1 (NF1)
    - Succinate dehydrogenase (SDH) deficiency

Understanding these classifications is essential for determining the appropriate diagnostic and treatment pathways.

Diagnosis of Pheochromocytoma

The Endocrine Society guidelines emphasize a multi-faceted approach to diagnosing pheochromocytoma. The diagnosis typically involves biochemical testing, imaging studies, and, in some cases, genetic testing.

Biochemical Testing

Biochemical testing is the cornerstone of pheochromocytoma diagnosis. The following tests are recommended:

1. **Measurement of Plasma Free Metanephrines:** Elevated levels of free metanephrines (metanephrine and normetanephrine) in plasma are highly indicative of pheochromocytoma.
2. **24-Hour Urinary Collection:** This test measures the total catecholamine and metanephrine levels over a 24-hour period, providing valuable diagnostic information.

3. **Plasma Catecholamine Levels:** While less commonly used, measuring plasma catecholamines can help confirm the diagnosis.

Imaging Studies

Once biochemical testing suggests pheochromocytoma, imaging studies are necessary to localize the tumor. The following imaging modalities are recommended:

- **Computed Tomography (CT) Scan:** Often the first-line imaging technique to identify adrenal tumors.
- **Magnetic Resonance Imaging (MRI):** Useful for patients with contraindications to CT or for assessing extra-adrenal lesions.
- **Positron Emission Tomography (PET):** Can be used in cases of metastatic disease or when other imaging methods are inconclusive.

Treatment Options

The mainstay of treatment for pheochromocytoma is surgical resection. However, preoperative management and postoperative care are critical components of the overall treatment strategy.

Preoperative Management

Before surgery, it is essential to control hypertension and manage symptoms to minimize the risk of intraoperative complications. The Endocrine Society guidelines recommend:

- **Alpha-Blockade:** Initiate treatment with an alpha-adrenergic antagonist to control hypertension. Phenoxybenzamine is often the drug of choice.
- **Beta-Blockade:** Initiate only after adequate alpha-blockade has been achieved to prevent worsening hypertension.
- **Volume Expansion:** Ensure adequate hydration to prevent intraoperative hypotension.

Surgical Resection

The primary treatment for localized pheochromocytoma is surgical excision. Laparoscopic adrenalectomy is the preferred approach in most cases due to its minimally invasive nature and quicker recovery time. The guidelines suggest:

- **Complete Tumor Resection:** Aim for complete removal of the tumor to reduce the risk of recurrence.
- **Assessment of Adrenal Function:** Monitor adrenal function postoperatively, especially if bilateral adrenalectomy is performed.

Postoperative Management and Follow-Up

Postoperative care is crucial for ensuring the best outcomes for patients with pheochromocytoma. The Endocrine Society guidelines highlight the importance of ongoing monitoring and management.

Postoperative Monitoring

Patients should be monitored for:

- **Hypertension:** Continuous blood pressure monitoring is necessary to manage any residual hypertension.
- **Adrenal Insufficiency:** Monitor for signs of adrenal insufficiency, particularly if the adrenal glands have been removed or damaged.
- **Complications:** Watch for complications such as infection or bleeding.

Long-Term Follow-Up

Long-term follow-up is essential for detecting recurrence or metastasis, especially in patients with hereditary syndromes. Recommendations include:

- **Regular Imaging:** Follow-up imaging studies may be warranted based on initial tumor characteristics.

- **Biochemical Monitoring:** Periodic measurements of plasma free metanephrines or urinary catecholamines.
- **Genetic Counseling:** For patients with hereditary syndromes, genetic counseling and testing for family members may be beneficial.

Conclusion

In summary, the **pheochromocytoma endocrine society guidelines** provide comprehensive recommendations for the diagnosis, treatment, and management of this complex condition. By following these guidelines, healthcare providers can improve patient outcomes through standardized care practices. Early diagnosis, effective preoperative management, surgical intervention, and diligent postoperative follow-up are all critical components in managing patients with pheochromocytoma. Understanding these guidelines is essential for clinicians to navigate the challenges presented by this rare neuroendocrine tumor.

Frequently Asked Questions

What is pheochromocytoma?

Pheochromocytoma is a rare tumor of the adrenal glands that produces excess catecholamines, leading to symptoms such as hypertension, palpitations, and sweating.

What are the key diagnostic criteria for pheochromocytoma according to the Endocrine Society guidelines?

The Endocrine Society guidelines recommend measuring plasma free metanephrines or 24-hour urinary fractionated metanephrines for diagnosis, along with imaging studies to confirm the presence of a tumor.

What imaging techniques are preferred for localizing pheochromocytoma?

The guidelines suggest using CT or MRI scans as the first-line imaging techniques to localize pheochromocytoma, with MIBG scintigraphy as an option for metastatic disease.

How should patients with pheochromocytoma be managed preoperatively?

Preoperative management includes controlling hypertension and minimizing cardiovascular risk, often achieved through the use of alpha-adrenergic blockers, with

beta-blockers added only after adequate alpha-blockade.

What is the recommended surgical approach for pheochromocytoma?

The guidelines recommend laparoscopic adrenalectomy as the preferred surgical approach for most patients with pheochromocytoma, unless contraindicated.

What follow-up care is suggested after surgical treatment of pheochromocytoma?

Postoperative follow-up should include monitoring blood pressure and plasma metanephrine levels to check for recurrence, typically within 3 to 6 months after surgery.

Are there any genetic considerations associated with pheochromocytoma?

Yes, the guidelines emphasize genetic screening for hereditary syndromes such as Multiple Endocrine Neoplasia (MEN) and Von Hippel-Lindau disease in patients diagnosed with pheochromocytoma.

What role do lifestyle modifications play in managing patients with pheochromocytoma?

While lifestyle modifications alone cannot treat pheochromocytoma, they can help manage symptoms and improve overall cardiovascular health in affected patients.

What are the potential complications of untreated pheochromocytoma?

Untreated pheochromocytoma can lead to severe complications, including hypertensive crises, cardiovascular disease, and increased mortality due to catecholamine excess.

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