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Fall 2014

# Diagnosis, Pathophysiology, and Clinical Treatment of a Patient with a Pheochromocytoma

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### Recommended Citation

Hale, Tricia L., "Diagnosis, Pathophysiology, and Clinical Treatment of a Patient with a Pheochromocytoma" (2014). *Nursing Student Class Projects (Formerly MSN)*. 59.  
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# Diagnosis, Pathophysiology, and Clinical Treatment of a Patient with a Pheochromocytoma

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## Introduction

Hypertension is an extremely common diagnosis today. This condition can be caused by a multitude of factors, and can inversely cause numerous unfavorable processes in the body. One rare but significant source of hypertension is a tumor known as a pheochromocytoma, or PCC.

This usually benign tumor is most often located on an adrenal gland, and can have a grave impact on the cardiovascular system. The tumor intermittently secretes an excess of the catecholamine norepinephrine, and if large enough, also secretes epinephrine [9]. These episodic bursts can cause extreme hypertension and tachycardia, leading to, at times fatal results. By understanding the pathophysiology and clinical presentation of a pheochromocytoma, this tumor can be considered as part of the early differential diagnoses by the advanced practice nurse. If a detailed history is obtained, the PCC patient can be treated appropriately and a potentially tragic outcome may be avoided. Surgical intervention is the most successful treatment modality, if the patient has been deemed a good candidate [3]. Careful monitoring must be performed pre-, during, and after surgery, noting any changes in vital signs or mental status. Pertinent patient education must be provided regarding significant symptoms, lifestyle modifications and when to seek immediate medical attention.

## Pathophysiology, Signs & Symptoms

Pheochromocytomas are tumors of the chromaffin cells, most commonly positioned on the adrenal medulla [8], although sometimes occur throughout the abdomen and thoracic regions, then known as paragangliomas [5]. Studies have indicated the majority of pheochromocytomas are idiopathic in nature, with one-fourth of diagnoses containing a genetic component. Both are usually diagnosed between the ages of thirty-five and fifty [5]. It is estimated that a PCC is present in four to five percent of the hypertensive population. They seem to occur equally between men and women [8]. They are challenging to diagnose, as they often appear dormant. When active, excessive catecholamine release causes a variety of symptoms, with hypertension as the most prominent. This hypertension occurs related to an increase in peripheral vascular resistance. Stressors to the body, such as exercise, caffeine intake, and surgery often prompt the catecholamine surge. Foods high in tyrosine, such as yogurt, wine and beer, can also spark their catecholamine release [9]. Effects of this hypertension, such as cardiomyopathy and infarction have been seen [1]. Palpitations, anxiety, and heat intolerance can occur. Fluctuation in blood levels of norepinephrine and epinephrine also lead to headaches and a potential decrease in cerebral blood flow [9]. One severe case has linked a PCC with a dissecting aneurysm of the vertebral artery [1]. Metabolic increases lead to diaphoresis and otherwise unexplained weight loss. Decreased insulin release can lead to hyperglycemia [9]. Because of the intermittent presentation of symptoms, As high as twenty-one percent of pheochromocytomas are without symptoms, which can be related to the patient's tolerance for high levels of catecholamines [8].



Radiologic image of a pheochromocytoma [10]

## Diagnosis

A diagnosis of PCC is made base on increased serum and urine catecholamine and catecholamine metabolite levels. Plasma free metenephrine measuring, done by immunoassay, has been declared more sensitive and accurate than measuring urine catecholamine levels. Positive levels are considered those over 90 ng/L and 125 nmol/day, respectively [6]. Unfortunately, many false-positive test results occur related to the low number of patients inflicted with a tumor of this sort. Elevated results also do not distinguish between the presence of a pheochromocytoma or a paraganglioma. Hence, once critical levels are identified, radiological studies, such as cat scan and magnetic resonance imaging, are then performed to confirm location and size [6].



Resected pheochromocytoma [7]

Although difficult to assess accurately, malignancy is suspected in up to ten percent of pheochromocytoma cases. Without metastasis, survival rate five years post-diagnosis is as high as eighty-nine percent. This percentage decreases drastically as metastasis is involved. Presently studies exist that are aimed at identifying genetic components and mutations, although on a large scale, these studies remain a challenge because the tumor is so rare [8].

## Treatment

Once diagnosis of a pheochromocytoma is made, options for treatment can then be discussed. Care is aimed at managing catecholamine excess and surges, in order to prevent hypertensive emergency and damaging cardiovascular effects. Alpha- and beta-adrenergic blockers are commonly prescribed. Titration of these vasoconstrictors is often performed on an inpatient unit for tighter control. Ultimately, if the patient is a candidate, surgical removal of the pheochromocytoma is the best treatment option. Surgery can be performed through a laparoscopic approach or by open incision adrenalectomy. Risk factors are reviewed to determine the most appropriate procedure. These include tumor size, patient age, body mass index, and known anesthesia complications [2]. Vital signs are closely monitored and controlled, before, during and after the procedure [3]. Surgery alone is often a cure, although chemotherapy may accompany the post-surgical regimen at times.

## Implications for Nursing Care

The informed nurse can be of great importance when diagnosing the cause of hypertension. By obtaining a detailed history of both chronic and intermittent symptoms, a pattern can be established. This pattern is vital when considering a diagnosis of a pheochromocytoma. Once diagnosed, the nurse must educate the patient regarding symptoms, emphasizing the importance of early recognition and when to seek immediate care. Stress reduction and lifestyle modification are important treatment modalities. Nurses need to teach their patients regarding the effect of weight management, diet and exercise, and non-smoking on healthier outcomes. Evaluating vital signs, medication effectiveness, reporting important findings to the physician, monitoring glucose levels, and assisting with pain control are all important nursing responsibilities, both prior to and following surgery.

## Conclusion

Although rare, a pheochromocytoma can lead to devastating, even fatal outcomes. Excessive epinephrine and norepinephrine surges often cause dangerous hypertension and cardiac decline. Understanding the pathophysiology involved will enable the advance practice nurse to consider this tumor as as part of the differential diagnoses. She can recognize symptoms, aid in diagnosing early, and monitor potentially life-threatening conditions closely. Treatment includes lab data collection, anti-hypertensive medication evaluation, monitoring of vital signs, and pre- and post-surgical care. Patient education regarding pheochromocytoma is key to patient success.

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