(UNDIAGNOSED EDITION)

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THE

PHEO VS FABULOUS

SURVIVAL GUIDE

A comprehensive e-book to help navigate the undiagnosed period with a rare disease called pheochromocytoma brought to you by Miranda Edwards the fabulous voice behind 'PHEO VS FABULOUS'

YOUR SURVIVAL GUIDE

YOUR SURVIVAL GUIDE

ABOUT THE AUTHOR

PHEO VS FABULOUS

My name is Miranda, the voice of *pheo vs fabulous.* I was first diagnosed with pheochromocytoma in 2010, I was only 19 years old.

After my surgery, my symptoms never got better. 4 years of ‘normal labs’ and hopeless appointments, I was found metastatic and terminal at 23 years old.

That was 11 years ago.

I’ve spent my life learning how to survive rare disease. Through surgeries, treatments, clinical trials, and many major setbacks.

I started www.pheovsfabulous.com so no one would have to go through what I did: being dismissed, misdiagnosed, and made to feel invisible.

I learned how to fight for every step, and now I’ve turned my survival into *your* blueprint.

It’s always been about one thing: ***helping you find your voice*** and remember who you are.

To my husband and co-Thriver ‘doctor cupcakes’ - thank you for helping me find the strength in my own voice.

This guide is free because *everyone* deserves access to information that could change their life.

While this is not medical advice, it is a carefully researched resource built from lived experience and years of advocacy.

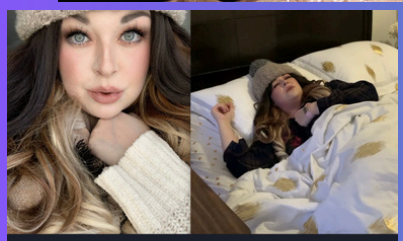
My hope is that it helps you ask better questions, advocate for yourself, and feel less alone.

If you'd like to support my ongoing cancer care, donations are appreciated but never expected.

You deserve answers. You deserve to be heard.

DONATE ❤

Miranda's advocacy has been featured in many publications and online media



Courtesy of Miranda Edwards

HEALTH Mar 29, 2023, 5:30 AM EDT

Doctors dismissed a young woman's heart-pounding adrenaline attacks as anxiety. She now has a deadly tumor.

Courtesy of Miranda Edwards

HEALTH Apr 6, 2023, 6:25 AM EDT

A woman with a terminal disease says she's still able to thrive despite her incurable condition. Here are her 3 pieces of advice.

2021-11-12

Not Just Surviving, But Thriving With Pheo vs. Fabulous

In this episode, we speak with Miranda Edwards, the voice behind Pheo vs. Fabu...

Cancer

Am I Ready to Accept That My Health Will Keep Declining?

Chronic Illness

What I Want You to Remember If You've Seen Relationships Fade After Becoming Sick

MONTRÉAL enSANTÉ

Health Monitor NETWORK

The MIGHTY

INSIDER

OUR health

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Fighting Pretty BLOG

HOS

BETWEEN BRC BOTH CHEEKS

A BRYGHT PRODUCTION

WAIT HOW DO YOU SPELL THAT?

A RARE DISEASE PODCAST

EMPOWERING YOU THROUGH DIAGNOSIS

You're likely here because you've been experiencing episodic symptoms that are hard to explain to your provider. I am here to empower you with INFORMATION. You're not alone anymore. We will do this together

LET'S GET STARTED

You're doing the best you can with the information you have right now...

It's hard to believe, but I know exactly what you're feeling right now. You fear no one will ever figure out what's wrong with you. You can't explain your symptoms because you look well when in front of your doctor.
I wrote this guide to empower you with information. So you can attend your appointment and know what to say! This survival guide will teach you how to fight for your life without using up your energy scrambling for information. This is your blueprint. A guide I once desperately needed - I created for all of us. Try saying this anytime you feel you need it: "everything is always working out for me no matter how it looks at any point in time" Remember, you're doing your best. Now it's time to give you all the information you need to be absolutely PHEARLESS.

WHAT ARE THE SYMPTOMS?

SIGNS & SYMPTOMS: PHEOCHROMOCYTOMA

When the tumor releases its high amount of adrenaline into the body, it causes a severe and sudden crisis and this often results in a spike of blood pressure and heart rate. The symptoms this causes may include:

- Severe onset headache
- visual disturbance
- heart palpitations
- chest pain
- nausea
- vomiting
- severe pain in the body/eyes/head/joints
- shaking
- sweating
- tremor
- chest pain resembling heart attack
- pallor
- impending doom

Symptoms vary from patient to patient. Some may experience more or much less, but most experience them in one large adrenaline crisis or (attack)

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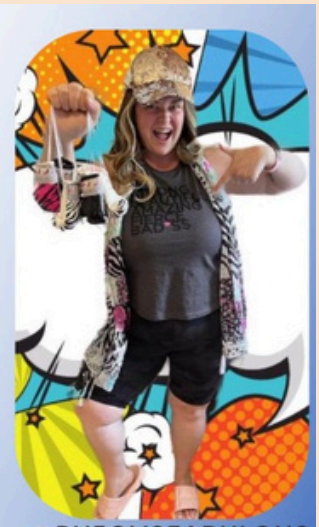

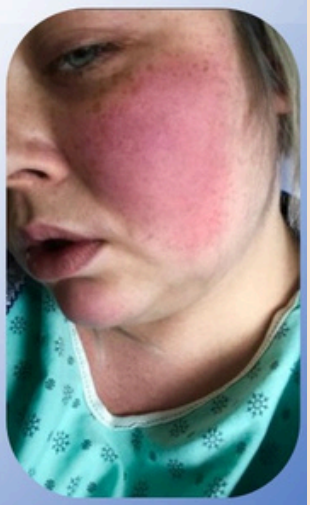

Do not rule out a pheo para because it's not a text book presentation. Many patients present with normal or even low blood pressure. You won't always have every symptom. Pheo para is called the great mimic because symptoms are similar to many other conditions. What separates it is the aggressive episodic nature of the symptoms.

Pheo Attack TRIGGERS

Although these attacks will happen randomly, there are also triggers that may make them more frequent or severe:

Exercise, movement, elevated HR

Heat, showers, sun exposure

Foods high in tyramine:

aged cheese, fermented and pickled foods (soy, miso, soy sauce) *alcohol* cured and processed meats (pepperoni, salami, sausage) citrus and tropical fruits: banana, avocado, pineapple, lemon, lime, orange, etc

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FOODS TO AVOID:

•Processed meats and Fish: this includes aged, cured, smoked, fermented, marinated, or preserved with nitrates and nitrites.

hot dogs, deli meat, sausage, bacon and ham are amongst the highest examples.

• Canned or prepared foods: canned soup, frozen dinners, lasagna, etc

• Foods that are high in yeast, even when homemade. *Especially* when homemade

•Beans contain high levels of tyramine due to the protein breakdown. Miso, tofu, and soy sauce contain the highest tyramine levels.

•Aged cheese: any cheese that is aged is going to present high. Safe cheeses are ricotta and cream cheese as these are not aged

•Vegetables and fruits: Overripe bananas, spinach and sauerkraut can be high in tyramine.

•Leftover food: to avoid tyramine spikes one should be eating fresh prepared food or freezing left overs and eating fresh

• Alcohol: Beer, ale, wines, whiskey and some liquors contain higher levels of tyramine. Gin and vodka may be safer alternatives but best to avoid all alcohol with pheochromocytomas

PHEO ATTACK RELIEF GUIDE:

I can only assume you are now on the appropriate combination of alpha and beta blockers for your blood pressure and heart rate. This will help in controlling the severity of episodes, but it won't stop them. These tips can be helpful while experiencing a 'pheo attack' otherwise known as a catecholamine crisis.

The following is a list of pheo safe recommended actions:

- Have water on hand to swallow pills
- Take an anti-nausea pill such as Zofran
- Consume an anti-anxiety prescription pill
- Elevate the legs into 'zero gravity'
- Have everyone speak quiet & calm
- Follow a deep breathing meditation
- Dim the lights or turn them off
- Apply a cold cloth or ice packs
- Rest rest rest in the aftermath

It's important for loved one's to remain calm. It's extremely uncomfortable for us. Hearing panic and loud voices makes the stress hormones even worse. Nothing can stop a catecholamine crisis, only the surgery to remove the tumor can offer that relief

These steps have been instrumental in keeping me safe during my attacks. I hope you feel the same.

FIND A DOCTOR RESOURCE:

Click here to find a doctor

An amazing resource by the carcinoid foundation allows for patients to search base on geographical preference. It shows which doctors are in your are only related to NET cancers and shows which are specialists

EXAMPLE:

Canada

Walter Kocha, MD (Oncology)
London, Ontario - SPECIALIST

David M. Liu, MD (Interventional Radiology)
Vancouver, British Columbia - SPECIALIST

Richard Malthaner, MD (Surgical Oncology)
London, Ontario

Peter Metrakos, MD (General Surgery and Transp Surgery)
Montreal, Quebec

Janice Pasieka, MD, FRCSC, FACS (Surgical Oncol General Surgery)
Calgary, Alberta - SPECIALIST

Juan Rivera, MD, FACE (Endocrinology and Metabolism)
Montreal, Ontario

Simron Singh, MD (Medical Oncology)
Toronto, Ontario - SPECIALIST

Florida

Shanel Bhagwandin, DO, MPH (Surgical Oncology)
Jupiter - SPECIALIST

Mark Bloomston, MD (Surgical Oncology)
Fort Myers

Aman Chauhan, MD (Medical oncology)
Miami - SPECIALIST

Seza A. Gulec, MD, FACS (Surgical Oncology/Radiology/Nuclear Medicine)
Miami - SPECIALIST

Boris G. Naraev, MD, PhD, FACP (Medical Oncology
Jupiter - SPECIALIST

Wasif M. Saif, MD (Hematology/Oncology)
Orlando - SPECIALIST

Jason S. Starr (Medical Oncology)
Jacksonville - SPECIALIST

Jonathan R. Strosberg, MD (Medical Oncology)
Tampa - SPECIALIST

From the onset of symptoms to a diagnosis can be long. This handbook will help at every step all in one place.

Now, we have to talk testing. You may be feeling dismissed right now, or like they won't consider the probability of it being such a rare condition. That's okay, what we need to focus on right now is evidence.

The next pages are going to be focused on biochemical testing. Things like blood and urine collections to start getting answers. With rare disease, it's not always straight forward. We have to ensure the correct lab protocols are followed for the samples to be viable. I've created and sourced different resources in order to print and take with you to the lab. This can be a challenging process, but sometimes goes smooth. This will make it easier for you to advocate for the results you need.

You're already worlds ahead of the herd!

BIOCHEMICAL TESTING

Plasma free metanephrines
(blood test)

(PPGL) tumors produce excess hormones called catecholamines, which are broken down to metanephrines. First line of action for diagnosis is to be tested at the onset of symptoms. Testing must be completed by a well informed lab, patient must be lying down in a supine position. Sample must be put on ice and transported within 30 minutes.

Avoid eating foods rich in tyramine (refer to foods to avoid list)

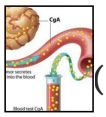
Please avoid alcohol – smoking – caffeine – exercise for a couple days prior

24hr urine fractionated catecholamines
(urine collection over 24hrs)

This measures the breakdown of adrenaline hormones in your urine. Both plasma and urine should be tested as per the endocrine society guidelines to ensure a clear result.

please avoid the same foods above and ask your doctor if you'll need to temporarily stop a particular medication!

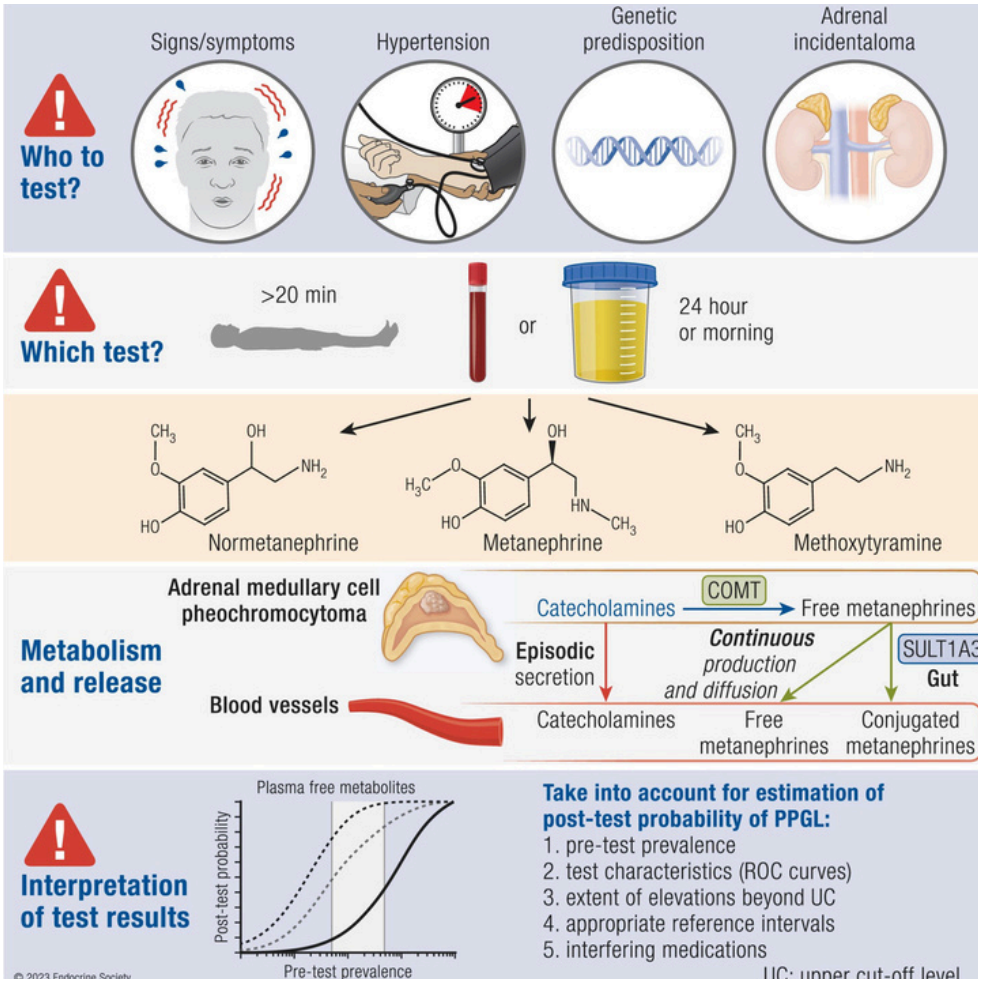
You will collect your urine for 24hours into a specimen collection jug. Specimen must be refrigerated and in a dark place, put on ice when transporting to the lab. (Extremely important) – once lab receives specimen, they too must refrigerate the sample or it will become invalid.



Chromogranin A
(Non specific NET cancer tumor marker)

Chromogranin A is a tumor marker for neuroendocrine tumors. In the absence of other elevated tests, it can help navigate a rare diagnosis and monitor the activity as it increases. Ask your doctor which medications need to be stopped before (ex) pantoloc

Infographic breakdown



Don’t forget 3-Methoxytyramine

3-methoxytyramine (3MT), is the breakdown of dopamine. This is a blood test that’s often forgotten! It can be useful in symptomatic patients to confirm a pheo para diagnosis. It’s also recommended for those who have a known genetic mutation such as SDHB.

Source: Biochemical Assessment of Pheochromocytoma and Paraganglioma Journal

NIH TESTING GUIDE:

1. INTERFERENCES. Acetaminophen interferes with the plasma metanephrine assay. It is therefore essential that the patient must not have taken acetaminophen in any form (e.g., Tylenol, Excedrin, combined cold medications) for at least 5 days before the sample is drawn. Several medications contain or are catechols and may interfere with the results of the catechol assay. These include alpha-methyldopa, alpha-methyl-para-tyrosine, isoproterenol, dobutamine, and carbidopa (such as in Sinemet™). In addition, dihydrocaffeic acid, a catechol metabolite of caffeic acid, can occur at high plasma concentrations in people who have recently ingested caffeinated or even decaffeinated coffee. For these reasons, we ask that all medications the patient is taking be listed on the accompanying information sheet. It is best if the sample be obtained in the morning after an overnight fast (water and non-caffeinated soft drinks are permissible).

2. BLOOD COLLECTION. The patient should be in the **supine position** for at least 20 minutes before and during collection of the blood sample. **To avoid the acute effects of the stress of venipuncture on plasma catechols and metanephrines it is essential that an indwelling i.v. be placed in a vein at least 20 minutes before the blood sample is drawn.** The sample may be drawn through an indwelling butterfly i.v. set or through an i.v. cannula (normal saline to keep the line patent), via a 3-way stopcock and Vacutainer adapter into a waste tube (saline void) and then into a chilled evacuated glass collection tube containing heparin in any form as an anticoagulant (e.g., a green top Vacutainer tube). It is preferable, but not essential, to draw the sample without a tourniquet.

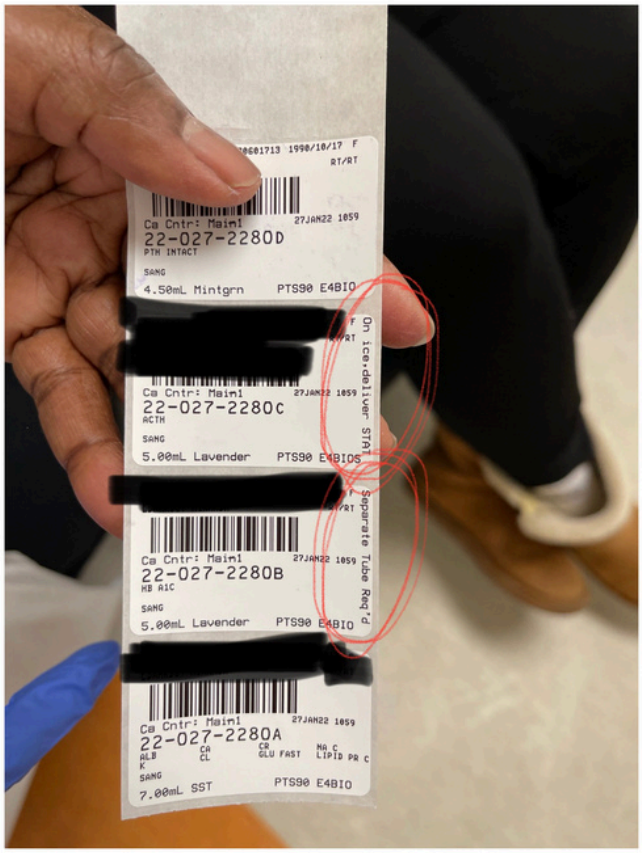
3. SAMPLE PROCESSING. The blood sample **(10 ml)** should be stored on ice until centrifuged (preferably at 4°C) to separate the plasma within 2 hours of blood collection. The plasma should be separated into **two aliquots**—one for catechols, one for metanephrines—with each aliquot at least 2 ml) in plastic tubes **clearly marked with the date and patient ID** and frozen immediately (e.g., on dry ice). The sample should be stored at -70°C or colder. Catechols in plasma stored at -20°C are not stable for extended periods, and so storage at -20°C should be limited to 2 weeks at most.

Plasma Metanephrines Guide:

**draw in prechilled tube, the blood sample should be kept on wet ice until centrifuged* also needs to be shipped refrigerated*

Storage/transport

Do not store. Send urgently to the laboratory on ice. Samples need to be separated and plasma frozen immediately.



Proper specimen collection for plasma metanephrines: pre chilled lavender tube delivered STAT on ice

BIOCHEMICAL TESTING

Plasma Metanephrines Guide:

For the lab: most patients wait an average of 4-5 years to get a rare diagnosis, please consider this with care when following this protocol. Thank you for the work you do and taking the time.

positive test results.(3) The recommended second-line test is measurement of fractionated 24-hour urinary metanephrines (METAF / Metanephrines, Fractionated, 24 Hour, Urine). In most cases this strategy will suffice in confirming or excluding the diagnosis. Occasionally, it will be necessary to extend this approach if there is a very high clinical index of suspicion or if test results are nonconclusive. In these cases, repeat plasma and urinary metanephrines testing, additional measurement of plasma or urinary catecholamines, or imaging procedures might be indicated.

Elevated results are reported with appropriate comments.

AA mayocliniclabs.com

BIOCHEMICAL TESTING

Pheo Para Tumor Imaging:

Once biochemical testing has confirmed levels of at least 2x or more, more diagnostics will follow. 4x fold elevation for diagnosis. However, the NIH considers 2x enough to investigate further. To begin, your doctor will typically order a CT or MRI scan.

Generally we see two different forms of imaging categorized into structural or functional. Structural could be an x-ray, CT, CAT scan or MRI.

Functional is looking at measuring the biological process, it's studying the function of the system. Normally a radioactive injection would accompany different types of functional or nuclear imaging such as:

123I-MIBG
18F-FDG-PET/CT
18F-DOPA-PET/CT
68Ga-DOTATATE PET/CT

The purpose of imaging is to take 'pictures' of inside the body. Imaging will confirm the location, the size, and if there's more than one tumor. It will also give information to what type of tumor it is, but unlike other tumors, pheo paras CANNOT be biopsied. If there is any suspicion at all of it being a pheo para, you should refuse this test and follow up with a specialist such as the NIH research institute or refer to the 'find a doctor' resource here

My tests came back negative, that means I'm all clear... right?

Pheo para diagnostics are difficult, and can be unclear.

We can't ignore the severity and episodic nature of these symptoms.

Here are some reasons the tests came back normal:

- improper lab testing and protocols
- protocols not followed correctly
- small tumors
- low level secretion from tumors
- non catecholamine secreting tumors

So, what do you do now?

Your doctor could repeat the testing or go ahead and do imaging such as CT or MRI to investigate further

Eyes to thighs imaging is recommended as these tumors can also be located in other regions which would then be called a paraganglioma


Advocacy phrases when you're not feeling heard
@pheovsfabulous
1
Can we revisit _____?
2
Could you please help me understand _____?
3
"I've been tracking this for weeks and it's only getting worse"
4
"This symptom is interfering with my ability to _____"
5
"I've tried _____ and_____ for x amount of time but have seen no change"
6
"could you help me better understand what YOU think it is?"

I'm not getting any answers, should I move on?

There are many factors involved here. The biggest one, you can't move on from the symptoms. They cannot be ignored, they're being caused by something.

pheopara ALLIANCE

Differential Diagnosis for Pheo

The clinical presentation of pheochromocytoma and paraganglioma can be highly variable, with similar signs and symptoms produced by numerous other clinical conditions. Therefore the tumor is often referred to as the "great mimic". Distinguishing pheochromocytoma from these and other less common conditions can be a challenge.

Endocrine
- ✓ hyperthyroidism, thyroid storm
- ✓ carcinoid
- ✓ hypoglycemia (often due to the presence of insulinoma)
- ✓ medullary thyroid carcinoma
- ✓ mastocytosis
- ✓ menopausal syndrome

Cardiovascular
- ✓ baroreflex failure

Neurologic
- ✓ migraine or cluster headaches
- ✓ diencephalic autonomic epilepsy
- ✓ meningioma
- ✓ POTS (postural orthostatic tachycardia syndrome)/ dysautonomia
- ✓ Guillain-Barre syndrome

Psychogenic
- ✓ anxiety or panic attacks
- ✓ factitious use of drugs
- ✓ hyperventilation

Pharmacologic
- ✓ tricyclic antidepressant
- ✓ cocaine
- ✓ alcohol withdrawal
- ✓ drugs stimulating adrenergic receptors
- ✓ abrupt clonidine withdrawal
- ✓ ephedrine-containing drugs
- ✓ factitious use of various drugs including catecholamines

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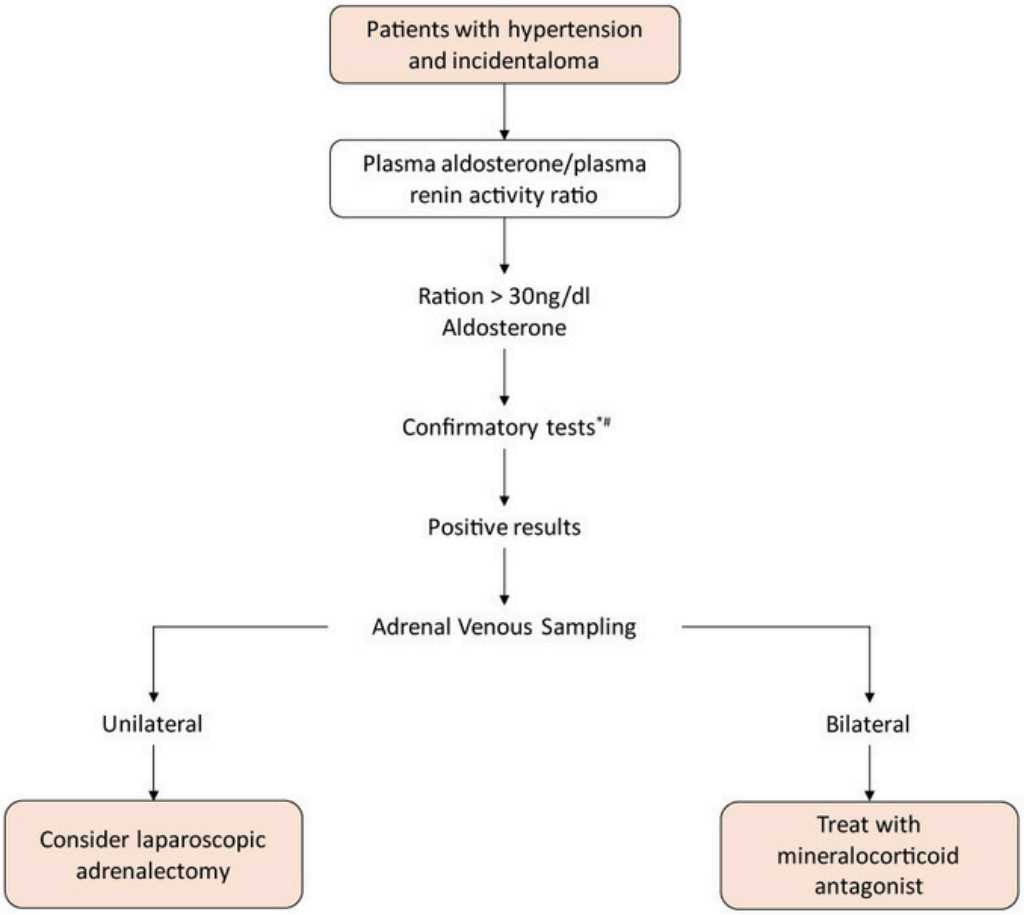
OTHER HORMONES TO TEST:

The symptoms can be similar to other conditions in the adrenal glands. This is a list of hormones that may be helpful for your GP. This is not a complete list, an endocrinologist referral will be helpful in your journey. As there are other types of tumors and nodules found on the adrenal glands

- **Aldosterone**
- **Plasma renin**
- **Cortisol**
- **Estrogen**
- **ACTH**
- **DHEA-S**
- **17-OH progesterone**
- **TSH**
- **Prolactin**
- **Testosterone**
- **IGF-1**
- **PTH**



ENDOCRINOLOGY TESTS

Not every answer comes quickly, and not every test gives the full picture — but every step you take is a piece of the puzzle. Even if this isn't the diagnosis, what you're doing matters. You are learning about your body, building a foundation for the care you deserve, and proving (to yourself and your team) that your voice matters. Stay curious, stay persistent, and remember: clarity is built one question, one test, and one fabulous step at a time.

For more resources, go to www.pheovsfabulous.com

Keep going my friend

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STAY FABULOUS WITH ME

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Nothing can take your fabulous

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