PRIMARY HYPERALDOSTERONISM

THE FACTS YOU NEED TO KNOW

NADF
National Adrenal Diseases Foundation

What is Primary Hyperaldosteronism?

Primary hyperaldosteronism is a disease caused by an excess production of the normal adrenal hormone aldosterone. This hormone is responsible for sodium and potassium balance, which then directly controls water balance to maintain appropriate blood pressure and blood volume.

What causes Primary Hyperaldosteronism?

People with a deficiency of aldosterone, especially found in association with cortisol deficiency in Addison's disease, have low blood volume and therefore low blood pressure, low sodium and high potassium. Just the opposite is seen in hyperaldosteronism. Primary hyperaldosteronism causes high blood pressure and a low serum potassium. The serum sodium is usually in the normal range. A separate disorder called secondary hyperaldosteronism is the result of abnormal fluid shifts seen in congestive heart failure, liver disease and certain kidney diseases.

Primary hyperaldosteronism used to be considered a rare disease, but recent studies show it to be a fairly common cause of hypertension. It is usually considered by physicians if they find an unexpectedly low potassium in a person being treated for hypertension. However, many people with this disease have potassium levels in the normal range. There is a growing awareness that many people with hypertension that is difficult to control or have progressively worsening hypertension may have primary hyperaldosteronism. While most individuals have no specific symptoms, some may have fatigue, headaches, muscle weakness and numbness. The physical examination is usually normal except for the elevated blood pressure.

There are two major types of primary hyperaldosteronism and three rare forms.

- **HYPERPLASIA** in both adrenal glands (about 60% of cases) is the most common cause.
- A **BENIGN TUMOR** of one of the adrenal glands is the cause in 35%. When the cause is a single adrenal tumor, it is labeled Conn's Syndrome.

A malignant tumor is a very rare cause. Other rare forms are unilateral hyperplasia and a familial glucocorticoid remediable form.

How is Primary Hyperaldosteronism diagnosed?

When primary hyperaldosteronism is considered by a physician, tests are done to look for an excess of the hormone aldosterone in the blood and urine, and also a suppressed plasma renin. The initial screening test is the aldosterone/renin ratio. To be certain that the value is accurate, serum potassium must be corrected first, and blood pressure medications that can affect aldosterone or renin must be discontinued for a few weeks. Borderline test results are common, so repetition of the ratio is often needed.

Other tests, looking at other adrenal steroid hormones, can be very useful as well as tests looking for the normal physiologic changes in hormones in the morning and evening, as well as responses to sodium challenge or sodium restriction.

Once primary hyperaldosteronism is confirmed, it is important to try to differentiate a single adenoma from hyperplasia. A CT of the adrenals will often show a small tumor in one of the adrenals. Typically, the adrenal tumors that cause hyperaldosteronism are very small, usually less than 2 cm. The presence of a tumor on CT is not adequate proof of an adenoma as the cause of the disease. Often it is misleading, with a tiny adenoma on the other side and a non-functioning tumor that is seen on the CT.

The gold standard for confirming unilateral disease is the use of adrenal vein sampling. This is a special procedure performed in major medical centers with experienced radiologists. Catheters are inserted in both adrenal veins and blood samples are obtained before and after ACTH infusion looking for a significant difference in levels of aldosterone between the two sides. If there is a significant difference, an adenoma is confirmed. Unfortunately, this procedure is not always diagnostic because it is difficult to get a catheter in the right adrenal vein.

How is Primary Hyperaldosteronism treated?

The treatment of primary hyperaldosteronism depends on the cause. If there is a single tumor, surgical removal of that tumor can cure the disease. The remaining adrenal gland is usually normal and individuals with this form of the disease will have enough adrenal hormone production from the remain-ing gland to live normally. Unfortunately, quite often there is still some residual hypertension even after the surgery, so sometimes antihypertensive medication is still necessary.

If bilateral hyperplasia is the cause of hyper-aldosteronism, or if surgery for an adenoma is not appropriate or safe, the disorder is treated with specific medications that block the effect of aldosterone. There are three such medications: spironolactone (Aldactone), eplerenone (Inspra) and amiloride (Midamor). These medications are very effective, but it is common to need to combine them with other antihypertensive medications for the management of hypertension in individuals with hyperaldosteronism.

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The National Adrenal Diseases Foundation is a non-profit organization providing information, education and support to all persons affected by adrenal disease. For more information on joining NADF, or to find a support group in your area, contact:

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