

Aldosterone-producing adenoma

Description

An aldosterone-producing adenoma is a noncancerous (benign) tumor that develops in an adrenal gland, which is a small hormone-producing gland located on top of each kidney. In most cases, individuals develop a single tumor in one of the adrenal glands. The adrenal tumor produces too much of the hormone aldosterone, which is a condition known as primary hyperaldosteronism. Aldosterone helps regulate the body's fluid levels and blood pressure by controlling the amount of salt retained by the kidneys. Excess aldosterone causes the kidneys to retain more salt than normal, which increases the body's fluid levels and blood pressure. People with an aldosterone-producing adenoma may develop severe high blood pressure (hypertension), and they have an increased risk of heart attack, stroke, or an irregular heart beat (atrial fibrillation).

Frequency

Aldosterone-producing adenomas cause up to 60 percent of cases of primary hyperaldosteronism. It is estimated that primary hyperaldosteronism accounts for 5 to 15 percent of cases of hypertension, which affects approximately 3 in 10 adults worldwide. However, the prevalence of aldosterone-producing adenomas is unknown.

Causes

Aldosterone-producing adenomas are caused by mutations in one of several genes. The most commonly mutated gene is *KCNJ5*, accounting for an estimated 40 percent of the tumors, followed by the *CACNA1D* and *ATP1A1* genes, which are mutated in about 9 percent and 6 percent of aldosterone-producing adenomas, respectively. Changes in other genes cause a small percentage of cases. Only about 60 percent of affected individuals have a mutation in one of the identified genes; additional unidentified genes are also thought to be involved in the condition.

The genes known to be involved in aldosterone-producing adenomas have roles in balancing the amounts of positively charged atoms (ions) of sodium (Na^+), potassium (K^+), and calcium (Ca^{++}) in cells. Each of the proteins produced from these genes transports certain ions across cell membranes. The flow of these ions creates an electrical charge across the cell membrane, which affects certain biochemical processes. In adrenal gland cells, this flow of ions helps control the production of aldosterone. Mutations in the *KCNJ5*, *CACNA1D*, or *ATP1A1* gene lead to abnormal electrical charges across cell

membranes. These abnormalities overactivate a biochemical process that increases adrenal cell growth and division (proliferation), which promotes adenoma formation. Overactivation of this biochemical process also increases aldosterone production, resulting in hyperaldosteronism and leading to hypertension.

Learn more about the genes associated with Aldosterone-producing adenoma

- ATP1A1
- CACNA1D
- CTNNB1
- KCNJ5

Additional Information from NCBI Gene:

- ATP2B3

Inheritance

Aldosterone-producing adenomas are generally not inherited but arise from a mutation in the body's cells that occurs after conception. In particular, the alteration occurs in adrenal gland cells that give rise to the tumor. Such mutations are called somatic mutations.

Other Names for This Condition

- Aldosterone-secreting adenoma
- Aldosteronoma
- Conn adenoma
- Primary aldosteronism due to Conn adenoma

Additional Information & Resources

Genetic Testing Information

- Genetic Testing Registry: Aldosterone-producing adrenal cortex adenoma (<https://www.ncbi.nlm.nih.gov/gtr/conditions/C1706762/>)

Patient Support and Advocacy Resources

- National Organization for Rare Disorders (NORD) (<https://rarediseases.org/>)

Clinical Trials

- ClinicalTrials.gov ([https://clinicaltrials.gov/search?cond=%22Aldosterone-producing adenoma%22](https://clinicaltrials.gov/search?cond=%22Aldosterone-producing+adenoma%22))

Scientific Articles on PubMed

- PubMed (<https://pubmed.ncbi.nlm.nih.gov/?term=%28Adenoma%5BMAJR%5D%29+AND+%28aldosterone-producing+adenoma%5BTIAB%5D%29+AND+english%5Bla%5D+AND+human%5Bmh%5D+AND+%22last+1440+days%22%5Bdp%5D>)

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