*Conn’s syndrome or primary hyperaldosteronism*

Conn's syndrome is a disease of the adrenal glands involving excess production of a hormone, called aldosterone. Find out why or how you get Conn's syndrome and what is its course

*What is Conn's syndrome?*

Conn's syndrome is a disease of the adrenal glands involving excess production of a hormone, called aldosterone. Another name for the condition is primary hyperaldosteronism.

Conn's syndrome is important because it is a potentially curable cause of high blood pressure (hypertension)..

Some studies suggest that Conn's syndrome is rare (one new case in a million people each year). However, when detailed investigations have been performed in patients with hypertension (high blood pressure), up to 15 per cent may have this condition.

Why or how do I get Conn's syndrome and what is its course?

The excess secretion of the hormone aldosterone into the blood is from an abnormal adrenal gland or glands.

Two types of abnormality are seen: a benign tumour of one adrenal, called an adenoma or a general enlargement of both adrenals, called hyperplasia.

The underlying reasons for the development of an adenoma or hyperplasia are not known.

What are the symptoms?

High blood pressure (hypertension) is the main, and often the only, symptom.

Other symptoms may occur because high aldosterone levels in the blood act on the kidney to increase the loss of the mineral potassium in the urine.

This in turn may lead to a fall in blood potassium, resulting in tiredness, muscle weakness and passing of large volumes of urine (polyuria), especially at night (nocturia).

However, these symptoms are also found in many other conditions (for example, diabetes mellitus or hypercalcaemia) and do not, by themselves, establish a diagnosis of Conn's syndrome.

Also, many patients with proven Conn's syndrome do not have a low blood potassium level.

How is Conn's syndrome diagnosed?

Conn's syndrome should be suspected in all patients with high blood pressure.

Traditional teaching has been to limit investigation for Conn's syndrome to patients who have a low blood potassium, or in whom blood pressure which is moderate to severe (>160/110mmHg) or is difficult to control with medication.

However, using these criteria, many patients with Conn's syndrome will not be diagnosed. For example, about 40 per cent of patients with proven Conn's syndrome have normal blood potassium levels.

The most rigorous method of diagnosis is to measure the blood levels of two hormones: aldosterone and renin (which plays a role in stimulating aldosterone production).

In Conn's syndrome, the aldosterone level is elevated and the renin level is low or undetectable…..so the aldersterone l renin ratio is the best screaning test.

What else could it be?

The commonest cause of high blood pressure is essential hypertension, and this may mimic Conn's syndrome.

Therefore high blood pressure and low blood potassium may be due to essential hypertension, which is being treated with diuretic drugs that cause a loss of potassium in the urine.

In addition, plasma renin activity may be suppressed by some drugs that are commonly used to treat hypertension (for example, beta-blockers). The suppression may mislead physicians to an incorrect diagnosis of primary hyperaldosteronism (Conn's syndrome).

There are also a few very rare conditions your doctor might need to exclude.

What can your doctor do?

Your family doctor can refer all patients with high blood pressure and low blood potassium to a specialist in endocrinology, renal disease or clinical pharmacology.

Other patients who should be referred for investigation include those with severe high blood pressure, or those who are poorly controlled on medication or who have a family member with an endocrine tumour.

However, there is a case for all patients with high blood pressure (>140/90mmHg) to have their aldosterone and plasma renin activity measured.

The difficulty is that the blood samples must be taken under controlled conditions, usually at 9am after lying down for 30 minutes, and require correct interpretation in the light of diet, drug therapy and other factors.

Therefore, in practice, aldosterone and plasma renin activity measurements are specialist investigations.

What can you do yourself?

A low salt diet may improve some symptoms by improving the low blood potassium and blood pressure.

However, lowering dietary salt intake may also cause an increase in the plasma renin activity (potentially masking the correct diagnosis).

So, for proper evaluation some patients may be asked to take a fixed intake of salt tablets for 72 hours before blood is taken for potassium, aldosterone and plasma renin tests.

What can your doctor do?

While awaiting evaluation at a specialist centre, potassium supplement tablets may be given to improve the low blood potassium. Some patients may have very low blood potassium, warranting urgent in-patient treatment.

Patients with Conn's syndrome will have to have special scans of the adrenal glands. This will allow the important distinction to be made between a single adenoma and hyperplasia of both adrenal glands. Blood may also be taken directly from the adrenal veins (via a catheter passed through a vein in the groin) to determine whether both adrenals are over-secreting aldosterone.

Treatment for an aldosterone-producing adenoma is surgical removal (unilateral adrenalectomy). This may be performed via laparoscopic (keyhole) approaches in some experienced centres.

Patients with bilateral hyperplasia (and also those with an adenoma awaiting surgery) may be treated with the drug spironolactone (eg Aldactone), which acts by blocking the effect of aldosterone. Spironolactone is somewhat similar in chemical structure to the female sex hormone, oestradiol, and therefore has some female hormone-like actions.

An alternative medication is called eplerenone (Inspra). Like spironolactone, this blocks the action of aldosterone.

Eplerenone is about half as strong as sprionolactone and usually has to be taken twice a day. The main advantage of eplerenone is that it does not have female hormone-like actions, such as increasing breast size or breast pain.

It's not licensed for use in Conn's syndrome (it is licensed for use in heart failure) – but nevertheless it's a useful treatment, especially for men with Conn's syndrome who have suffered breast side-effects from spironolactone.

Without drug or surgical treatment, high blood pressure in many patients with Conn's syndrome is difficult to control.

Poorly controlled high blood pressure is associated with increased rates of stroke, heart disease and kidney failure.

*Prognosis*

Treatment of Conn's syndrome is usually successful. Many patients with a single adrenal adenoma will be able to stop drug treatment and will have normal blood pressures.

Nevertheless, many specialist centres will follow a patient with Conn's syndrome for life. This is to monitor the rare possibility of growth of a second adenoma.

Patients with bilateral hyperplasia should have life-long monitoring of effectiveness and side-effects of drug treatment. Again, quality of life is generally good, although some patients may not be able to tolerate spironolactone treatment.?