CONDITION

Multiple endocrine neoplasia (MEN) I

An inherited condition that raises the risk of tumours in several hormone-producing glands.

WHAT IT IS
Inherited tendency to develop multiple hormone-producing tumours

BODY SYSTEM
Hormone-producing glands and digestive organs

MAIN SYMPTOMS
Kidney stones, stomach ulcers, low blood sugar, headaches

WHAT CAUSES IT
Inherited change in the MEN1 gene

HOW IT IS DIAGNOSED
Clinical criteria or MEN1 genetic testing, supported by imaging

MAIN TREATMENT
Lifelong surveillance, surgery, and hormone- or tumour-controlling medicines

CAN IT BE CURED?
No - inherited tumour risk remains lifelong

IS IT AN EMERGENCY?
Sometimes - confusion, seizure, or sudden vision loss

LONG-TERM OUTLOOK
Often manageable with lifelong surveillance and timely treatment

Overview

Multiple endocrine neoplasia type 1, usually shortened to MEN1, is a rare inherited condition. It increases the chance of developing tumours in several endocrine glands. These glands make hormones, which are chemical messengers that control body functions.

MEN1 mainly affects the parathyroid glands, the pituitary gland, and hormone-producing cells in the pancreas and first part of the small intestine. It can also cause tumours in the adrenal glands, lungs, thymus, skin, and other tissues.

Many MEN1 tumours are benign, meaning they are not cancer. Even so, they may make too much hormone or press on nearby structures. Some neuroendocrine tumours can become cancerous and spread.

MEN1 affects people differently, including members of the same family. One person may have few problems, while another develops several tumours over time. Regular monitoring matters because a tumour or hormone change may be found before it causes noticeable symptoms.

Symptoms

MEN1 may cause no symptoms at first. Symptoms depend on which glands are affected, whether a tumour releases hormones, and how large it becomes.

Overactive parathyroid glands cause primary hyperparathyroidism. This raises the calcium level in your blood. Possible effects include:

Pituitary tumours grow in the gland at the base of the brain. A tumour that releases prolactin may cause irregular or absent periods, breast milk production when not breastfeeding, reduced interest in sex, or erection problems. Other pituitary tumours can cause changes in growth, facial features, sweating, weight, or cortisol levels.

A larger pituitary tumour may cause headaches or loss of side vision. Sudden severe headache with rapid vision loss is an emergency.

Neuroendocrine tumours can develop in the pancreas or small intestine. Their symptoms depend on the hormone involved:

Some neuroendocrine tumours do not release enough hormone to cause symptoms. They may be found during routine scans or after they grow large enough to cause pain, blockage, or pressure.

Skin findings can include small reddish facial bumps called angiofibromas, firm collagen-containing bumps, and fatty lumps called lipomas. These are usually not harmful but may support the diagnosis.

- Kidney stones, thirst, or passing urine often
- Constipation, nausea, abdominal discomfort, or tiredness
- Muscle weakness, low mood, or difficulty concentrating
- Bone thinning or fractures over time
- Gastrin-producing tumours can cause repeated stomach ulcers, severe acid reflux, abdominal pain, or diarrhoea
- Insulin-producing tumours can cause low blood sugar, with sweating, shaking, hunger, confusion, or fainting

- Other functioning tumours may cause watery diarrhoea, a distinctive rash, weight loss, or changes in blood sugar

Causes and risk factors

MEN1 is caused by a harmful change, also called a pathogenic variant, in the MEN1 gene. This gene normally helps control cell growth by making a protein called menin. When it does not work properly, certain cells are more likely to form tumours.

The condition follows an autosomal dominant inheritance pattern. This means a parent with the genetic change has a 1 in 2 chance of passing it on in each pregnancy. MEN1 can affect people of any sex.

Sometimes the genetic change starts in a person for the first time, with no known family history. This is called a de novo variant. That person can still pass it to their children.

Having a close relative with MEN1 or a known MEN1 variant is the main risk factor. MEN1 is not caused by diet, stress, alcohol, or something a parent did during pregnancy.

Diagnosis

MEN1 may be diagnosed from its pattern of endocrine tumours, a family history, or genetic testing. The usual clinical pattern involves two or more characteristic tumours affecting the parathyroid glands, pituitary gland, or pancreatic and intestinal neuroendocrine cells.

A blood or saliva genetic test can look for a pathogenic variant in the MEN1 gene. Testing is especially informative when the specific family variant is already known. A genetics professional can explain the possible results and what they may mean for relatives.

A negative genetic test does not always settle the diagnosis. Some people meet clinical criteria without an identifiable MEN1 variant, and other inherited conditions can produce a similar pattern.

Further tests check which organs are affected. They may include:

Imaging helps locate and measure tumours. It does not replace hormone testing or genetic assessment. Because some MEN1-related tumours develop before symptoms begin, people with a confirmed diagnosis usually enter a lifelong surveillance programme.

- Blood tests for calcium, parathyroid hormone, pituitary hormones, glucose, and selected digestive hormones

- Urine tests for particular hormone changes
- MRI, CT, ultrasound, or specialised endoscopic ultrasound
- Functional imaging that shows receptors or activity in neuroendocrine tumour cells

Treatment and management

There is no single treatment that removes the inherited tendency to form tumours. Care focuses on finding changes early, controlling excess hormones, and treating individual tumours when needed.

Management is often coordinated by an endocrinology team. Depending on the organs involved, care may also include endocrine surgeons, gastroenterologists, oncologists, neurosurgeons, radiologists, and genetics professionals.

Overactive parathyroid glands may be treated with surgery. The operation is planned carefully because several glands are often affected. Calcium can fall after surgery, so blood tests and sometimes calcium or vitamin D replacement are needed. Parathyroid overactivity can return or persist, making long-term follow-up important.

Pituitary treatment depends on the hormone produced and the tumour's size. Prolactin-producing tumours often respond to medicines that reduce prolactin and shrink the tumour. Other pituitary tumours may require surgery, hormone-controlling medicines, or occasionally radiotherapy.

For pancreatic and intestinal neuroendocrine tumours, treatment may include:

An insulin-producing tumour may require surgery and measures to prevent low blood sugar while treatment is planned.

Not every small tumour needs immediate surgery. The team may monitor its size, growth, hormone activity, location, and signs of spread. This is active surveillance rather than a lack of treatment.

- Medicines to control stomach acid or other hormone effects
- Surgery for selected tumours
- Somatostatin-type medicines that can control hormones and tumour growth
- Targeted medicines, chemotherapy, or peptide receptor radionuclide therapy for selected advanced tumours
- Procedures aimed at tumours that have spread to the liver

Complications

High calcium over a long period can lead to kidney stones, reduced kidney function, and weaker bones. Very high calcium may cause dehydration, vomiting, marked drowsiness, or confusion.

Repeated low blood sugar can cause falls, accidents, seizures, or loss of consciousness. Severe acid production from a gastrin-producing tumour can lead to ulcers, bleeding, or a hole in the stomach or intestine.

Pituitary tumours may affect vision or reduce normal pituitary hormone production. Rarely, sudden bleeding into a pituitary tumour causes pituitary apoplexy, with a severe headache, vomiting, eye movement problems, or rapid vision loss.

Some pancreatic, intestinal, lung, or thymic neuroendocrine tumours can be cancerous and spread to lymph nodes, the liver, or other organs. The risk is not the same for every tumour. Its type, size, rate of growth, and stage all matter.

Treatment itself may cause complications, including low hormone levels after surgery. These can often be managed with monitoring and hormone replacement when appropriate.

Prevention

MEN1 itself cannot currently be prevented. A person is born with the genetic change, and no diet, supplement, or lifestyle measure has been shown to stop MEN1 tumours from developing.

Regular surveillance cannot remove the risk, but it may find hormone changes or tumours before they cause serious problems. Keeping scheduled blood tests, scans, and eye or bone assessments is therefore an important part of reducing harm.

Genetic counselling can help relatives understand testing. People planning a pregnancy can also discuss inheritance, testing options, and reproductive choices. These are personal decisions, and counselling should provide information without pressure.

Not smoking supports general health and may be particularly relevant when the chest is being monitored, but it does not prevent the underlying condition.

Living with it and what to expect

MEN1 is lifelong. New tumours can appear over months or years, and a previously treated problem may return. Surveillance usually continues even when you feel well and all recent tests

are normal.

The pattern is difficult to predict from the genetic result alone. Some people mainly need monitoring and treatment for excess calcium. Others need several operations or medicines for different tumours. Tumours that are growing, releasing hormones, or showing signs of spread usually require closer attention.

Many people continue with work, education, relationships, travel, and family life. Appointments, scans, medication, and recovery from surgery can still be disruptive. Asking whether tests can be coordinated through one specialist centre may make long-term care easier.

Keep an up-to-date list of diagnoses, operations, medicines, hormone replacement, and the family MEN1 variant if known. This information can be useful when seeing a new clinician or receiving emergency care.

Pregnancy may require advance planning because calcium levels, pituitary tumours, medicines, and imaging may need special review. Do not stop prescribed treatment without discussing it with the team overseeing your care.

Long-term outlook is shaped mainly by the types of tumour present, whether they can be controlled, and whether a cancerous neuroendocrine tumour has spread. Consistent follow-up and treatment have an important role, but no individual course can be guaranteed.

When to get medical help

Attend routine follow-up even when you have no symptoms. Contact your usual care team if you develop new kidney stones, persistent abdominal pain, repeated reflux or ulcers, unexplained diarrhoea, menstrual changes, nipple discharge, headaches, or changes in vision.

Seek prompt medical advice for repeated episodes of sweating, shaking, confusion, or unusual hunger, especially if they improve after eating. These may be signs of low blood sugar and need assessment.

Get urgent or emergency help for:

Do not drive yourself if you are confused, faint, having major vision changes, or may have severely low blood sugar.

- A seizure, collapse, loss of consciousness, or severe confusion
- A sudden severe headache with vision loss or eye movement problems
- Vomiting blood, black sticky stools, or severe sudden abdominal pain
- Severe weakness, repeated vomiting, dehydration, or marked drowsiness

- Symptoms of low blood sugar that do not improve quickly with available treatment

Common questions

Is MEN1 a type of cancer?

MEN1 is not itself cancer. It is an inherited condition that increases the chance of several tumours. Many of these tumours are benign, but some neuroendocrine tumours can become cancerous or spread.

Can MEN1 skip a generation?

A person who does not inherit the family MEN1 variant cannot usually pass that variant on. MEN1 can appear to skip a generation if a relative was never tested, had mild symptoms, or died before tumours were recognised.

What is the difference between MEN1 and MEN2?

They are separate inherited conditions caused by different genes. MEN1 commonly affects the parathyroid, pituitary, pancreas, and small intestine. MEN2 is linked to changes in the RET gene and has a different tumour pattern, including medullary thyroid cancer.

Can I have children if I have MEN1?

Yes. Each child of a parent with MEN1 has a 1 in 2 chance of inheriting the variant. A genetics professional can explain testing and reproductive options before or during pregnancy.

Does everyone with MEN1 need surgery?

No. Surgery is based on which gland is affected, hormone levels, symptoms, tumour size and growth, and the chance of cancer or spread. Some findings are monitored with regular tests and scans.

Is there a special MEN1 diet?

There is no diet that prevents MEN1 tumours. Your food choices may need adjustment for a specific problem, such as kidney stones, ulcers, low blood sugar, or recovery after treatment. Advice should match that problem rather than MEN1 as a whole.

Sources

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