# MULTIPLE ENDOCRINE NEOPLASIA TYPE 1


## Multiple Endocrine Neoplasia (MEN 1)

**Multiple Endocrine Neoplasia type 1 (MEN 1)** is a rare, inherited disorder that causes tumors in the parathyroid glands, pituitary gland, and the endocrine tissue of the gastro-entero-pancreatic (GEP) system.
While the most common issues are hyperparathyroidism, pancreatic neuroendocrine tumors, and pituitary adenomas, patients can also develop other non-endocrine tumors, such as lipomas, angiofibromas, and adrenal tumors.

### Pituitary

The pituitary gland is about the size of a pea or cherry, and hangs down from the main part of the brain. It regulates various body functions and plays an important role in controlling hormone levels in the body.

### Parathyroids

The parathyroid glands are two pairs of small, oval-shaped glands. They are located next to the two thyroid gland lobes in the neck. Each gland is usually about the size of a pea.

### Pancreas

The pancreas is a gland located in the upper abdomen that produces both hormones and digestive enzymes.
It lies horizontally in the upper abdomen, directly behind the stomach and in close proximity to the duodenum, spleen, liver, and major abdominal blood vessels.

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## Multiple endocrine neoplasia type 1: Recommendations and guidelines for best practice What is new compared to the current practice?

Because there is still a lot we don't know about the best ways to care for people with MEN-1, a group of the world's top specialists came together to share their knowledge. In a step-by-step, anonymous survey (called a Delphi study), 81 experts in three areas that MEN-1 affects—parathyroid glands, neuroendocrine tumours, and the pituitary gland—answered a series of questionnaires. After each round, the combined answers were sent back to the group for further discussion, allowing the doctors to refine their opinions until they reached a clear, agreed-upon set of recommendations. The final virtual consensus meeting produced 55 recommendations, each given a strength that shows how strongly the experts support it. These recommendations were published recently, and we have distilled the most important points for you on the following slides.

**Source: Brandi et al. Lancet Diabetes Endocrinol 2025 https://doi.org/10.1016/s2213-8587(25)00119-6**

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## Source for the Patients´ Summary

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Multiple endocrine neoplasia type 1 (MEN1): recommendations and guidelines for best practice

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## Compiled by

EMENA (www.emena.eu), the European Multiple Endocrine Neoplasia Alliance. EMENA is an umbrella organisation for patient groups and medical experts, who provide information and support services to patients with MEN and their relatives. EMENA was founded in 2013 and is a German not-for-profit organisation.

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EUROPEAN MULTIPLE ENDOCRINE NEOPLASIA ALLIANCE

# MULTIPLE ENDOCRINE NEOPLASIA TYPE 1

## Screening for asymptomatic MEN 1 adults

**Biochemistry (once a year)**
Calcium
Prolactin
IGF-1
Gastrin

**Imaging**

| | |
|---|---|
| MRI Abdomen | every 2-3 years |
| MRI Pituitary | every 3-5 years |
| CT Chest | every 3-5 years |

**It is explicitly recommended that all MEN patients in all age groups receive care at a specialist centre.**

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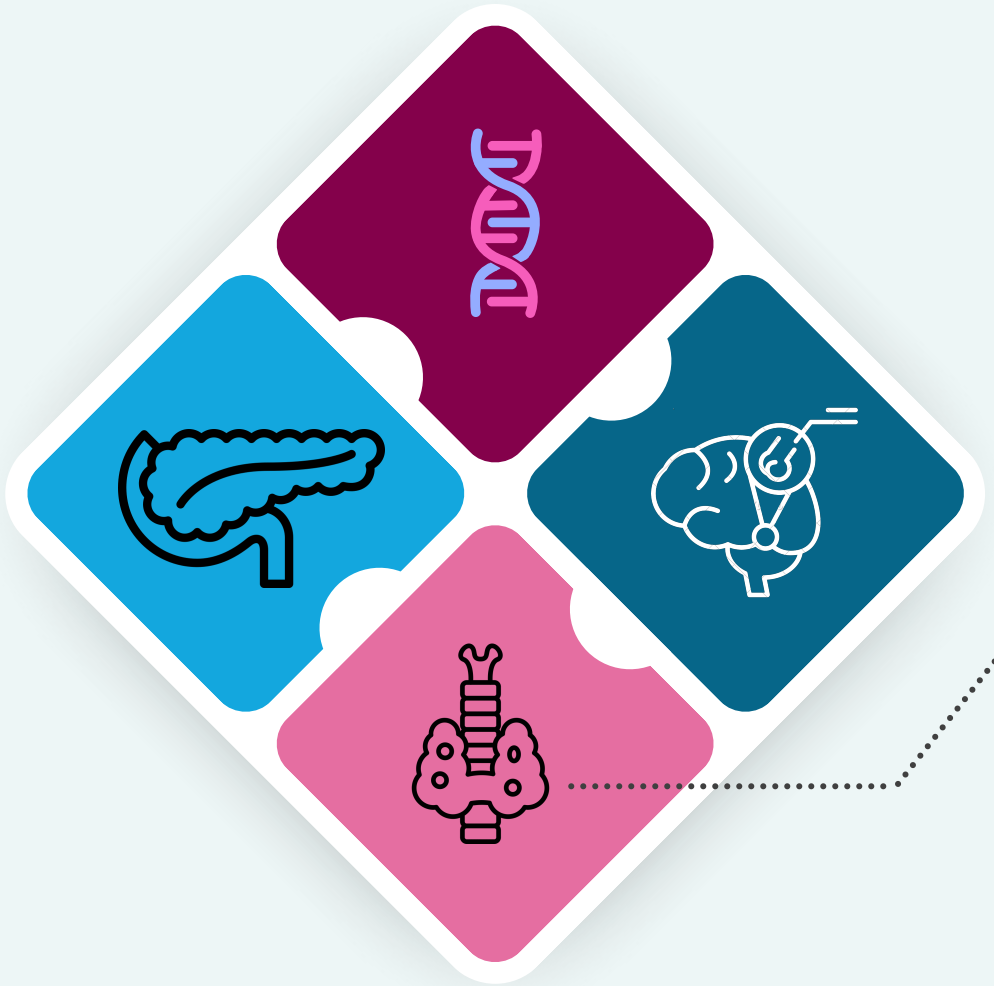

## Parathyroidectomy – Indication and timing

For the first time, the new recommendations now include several indications for surgery:

- symptomatic or target organ involvement
- Ca > 1mg/dL(0,25 nmol/l) above the upper limit of normal in adults and suggested in children, irrespective of symptoms
- take presence of gastrinoma in account considering timing.

Subtotal (3 – 3,5 gland) parathyroidectomy as initial surgery for children and adults.

Unilateral clearance is suggested only in selected cases.

A transcervical thymectomy is recommended.

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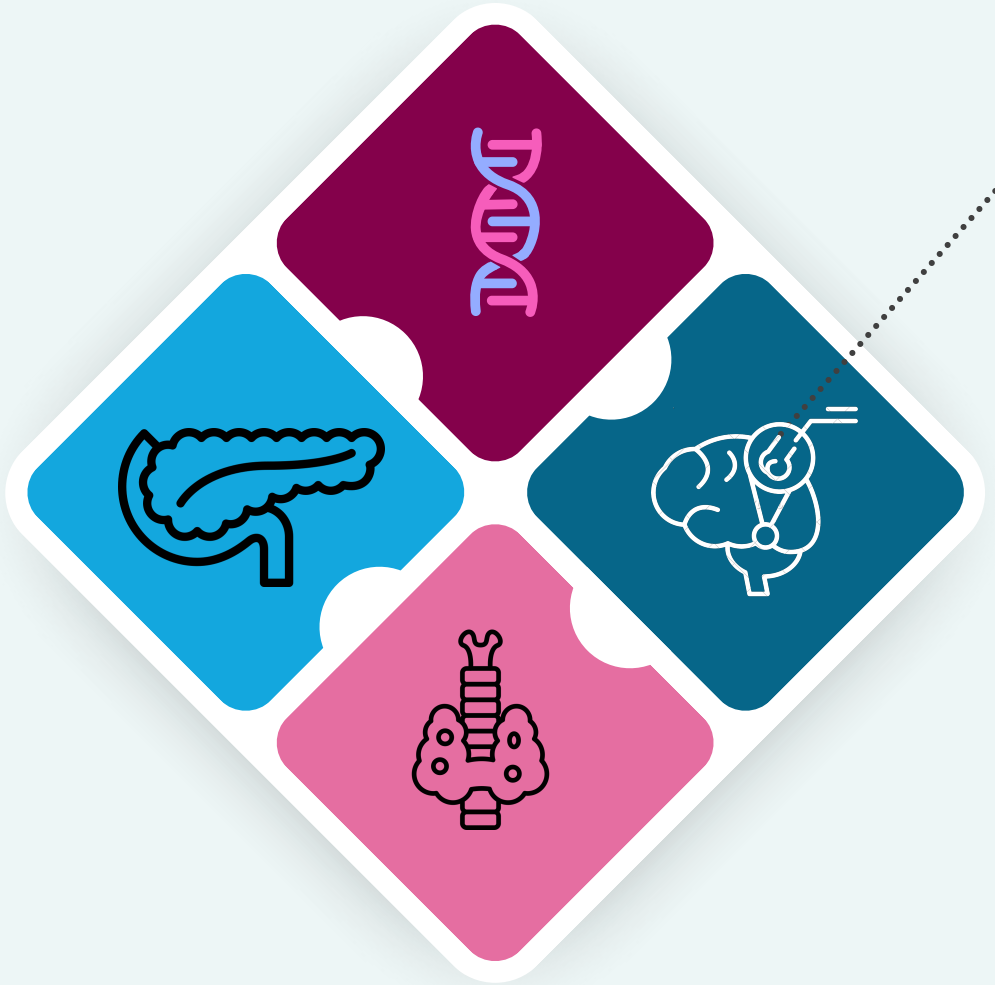

## Pituitary

**Screening every 3-5 years**
End of screening at age 75 years
Management similiar to sporadic adenomas

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## Pancreas

- Tumor ≤ 2 cm and growth < 1mm / year: Imaging every 1 -2 years
- Endoscopic ultrasound when results affect management
- no routine biopsy
- Somatostatin receptor PET-CT in patients planned for surgery only when results affect the management (suggested)
- Surgery for tumors > 2 cm and growing

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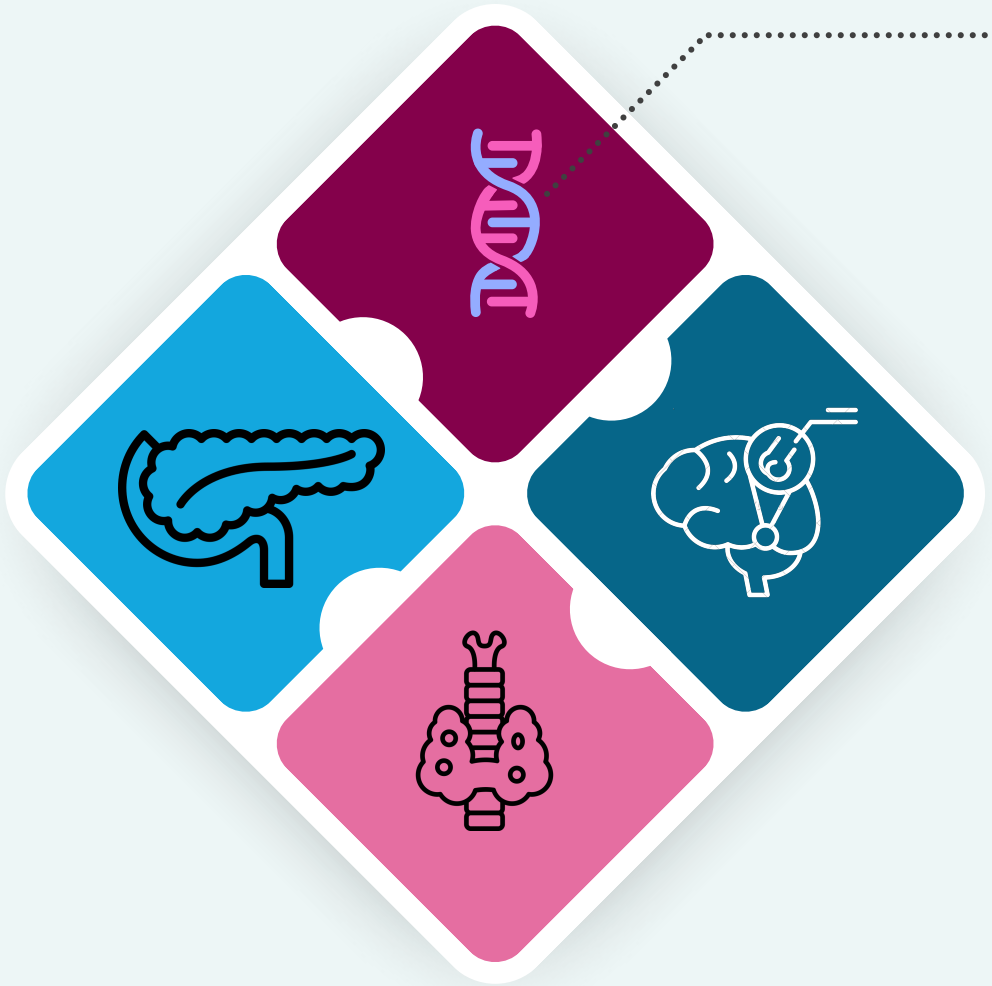

## Further manifestations

**Thoracic NET:**

Screening and management

- Initiation of screening at age 20–25
- Screening asymptomatic adults every 3 – 5 years (suggested)
- Lung – NET < 1 – 2 cm and growth < 1 mm/year: Imaging every 1 – 2 years

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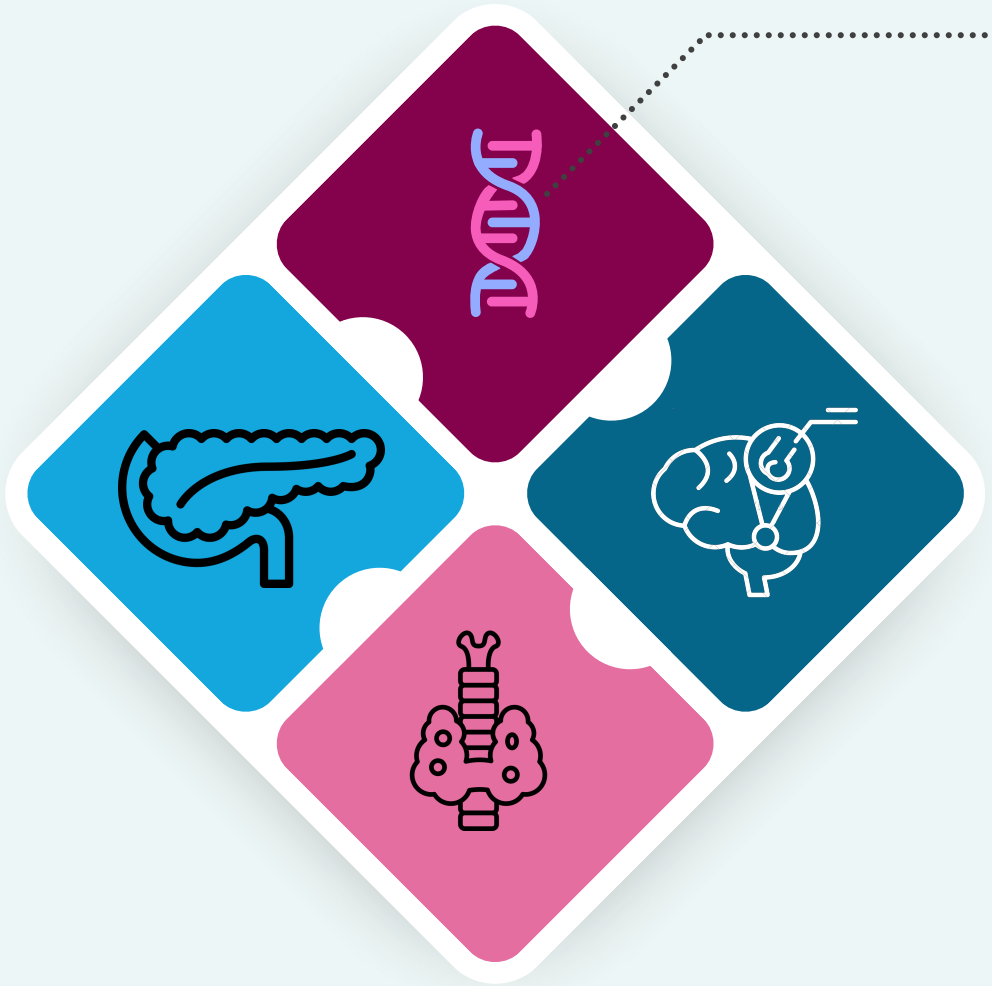

## Further manifestations

**Adrenal cortical tumours:**
Screening and management

- in most cases non-functioning, although hormonal hypersecretion possible (primary hyperaldosteronism (Conn´s syndrome) and ACTH-independent Cushing´s syndrome)
- Pheochromocytomas are extremely rare in MEN 1 patients.
- Functioning adrenal tumors secrete cortisol or/and sex steroids.
- Biochemical investigation for symptomatic adults or tumours >1 cm : Renin and Aldosterone
- no consenus for management of non-functioning adrenal adenomas.

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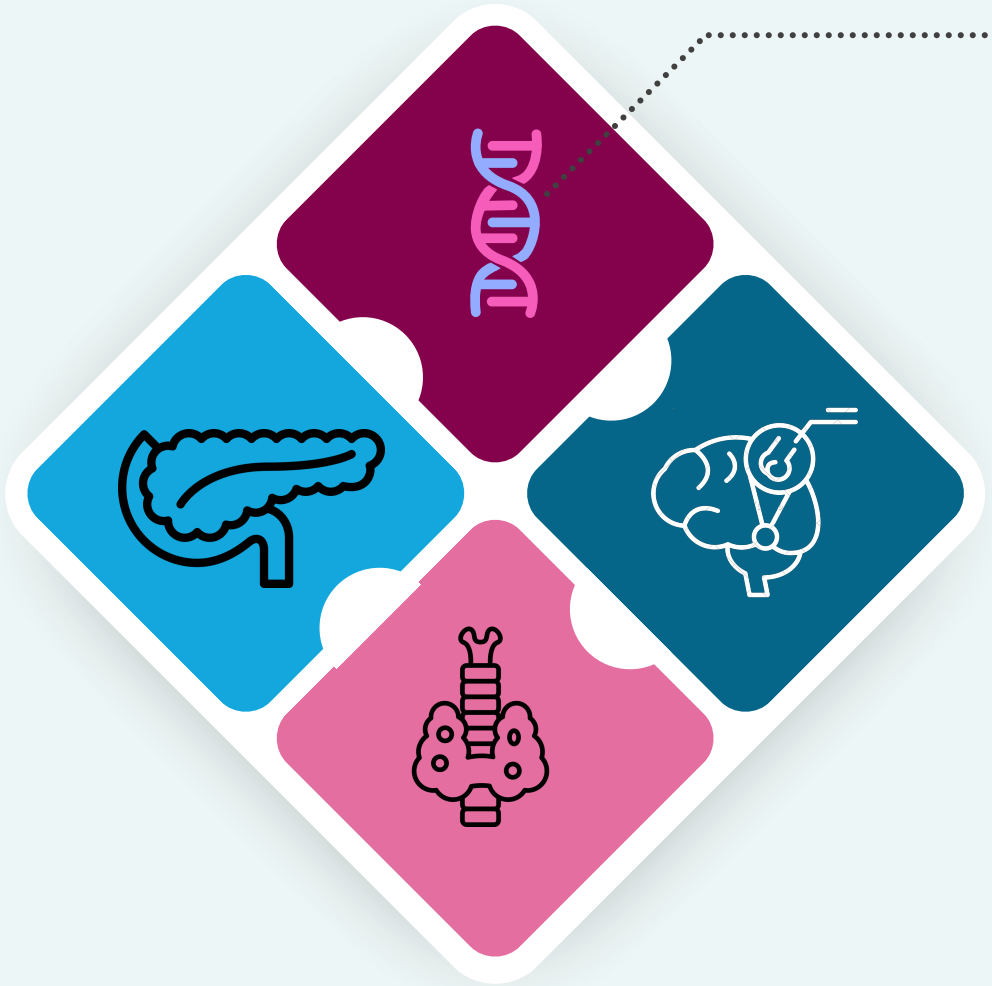

## Further manifestations

**Adrenal cortical tumours:**

Treatment

- Surgery for non-functioning tumours > 4 cm with atypical or suspicious radiological features and are 1-4 cm in diameter
  or
- show growth within a 6- month interval.
- treatment of functioning adrenal tumours is similiar to non-MEN 1 associated tumours.

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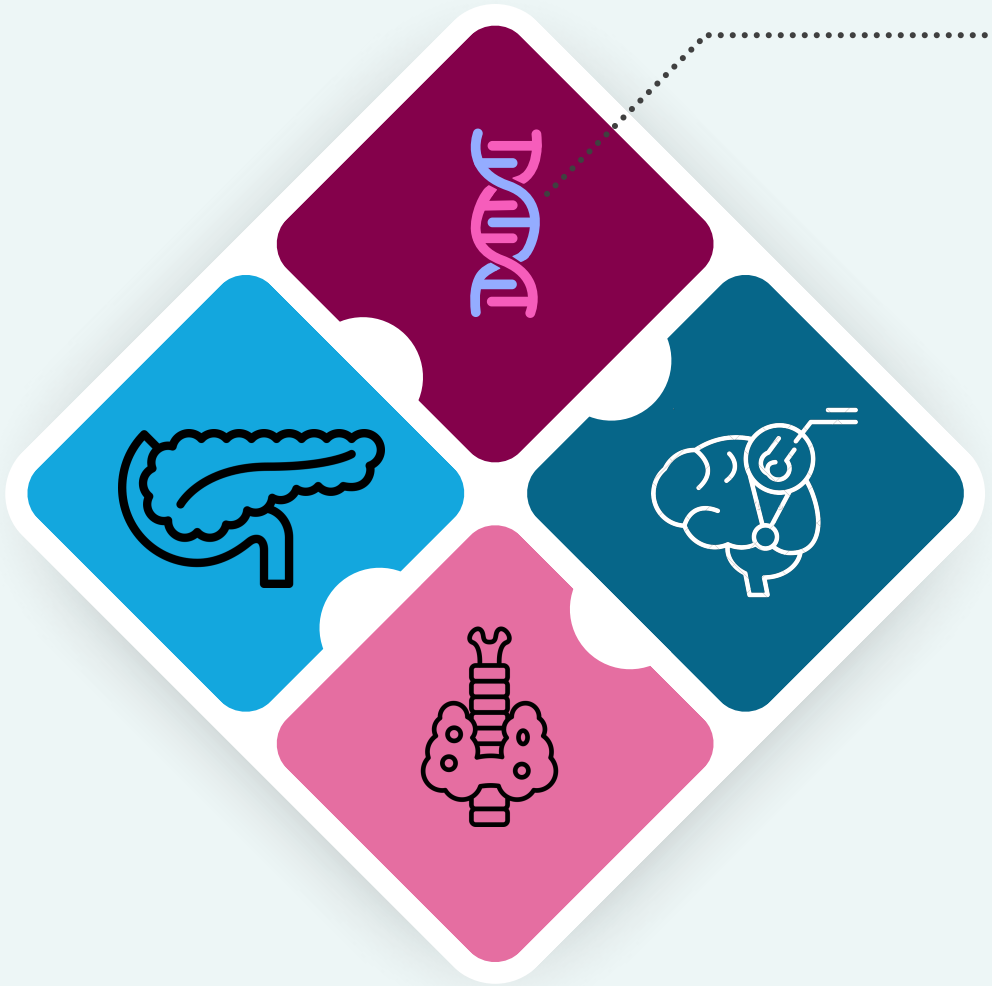

## Further manifestations

Some studies have shown that women with MEN 1 may have an increased risk of breast cancer. Currently, there is insufficient evidence to consider breast cancer as part of MEN 1.

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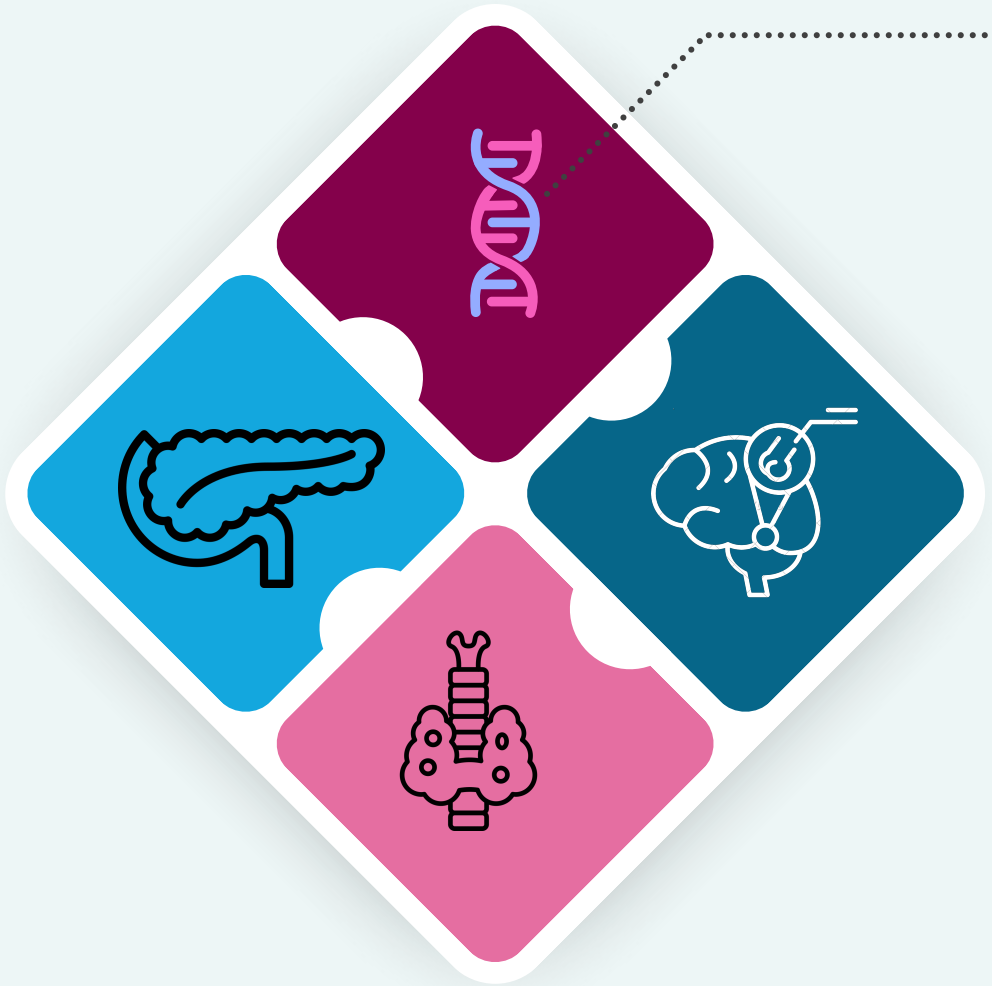

## Further manifestations

Patients with MEN 1 may develop angiofibromas and collagenomas of the skin, as well as lipomas. In addition, patients are at risk of developing leiomyomas, mainly in the upper gastrointestinal tract and urogenital area. Leiomyosarcomas or other sarcomas are rare.

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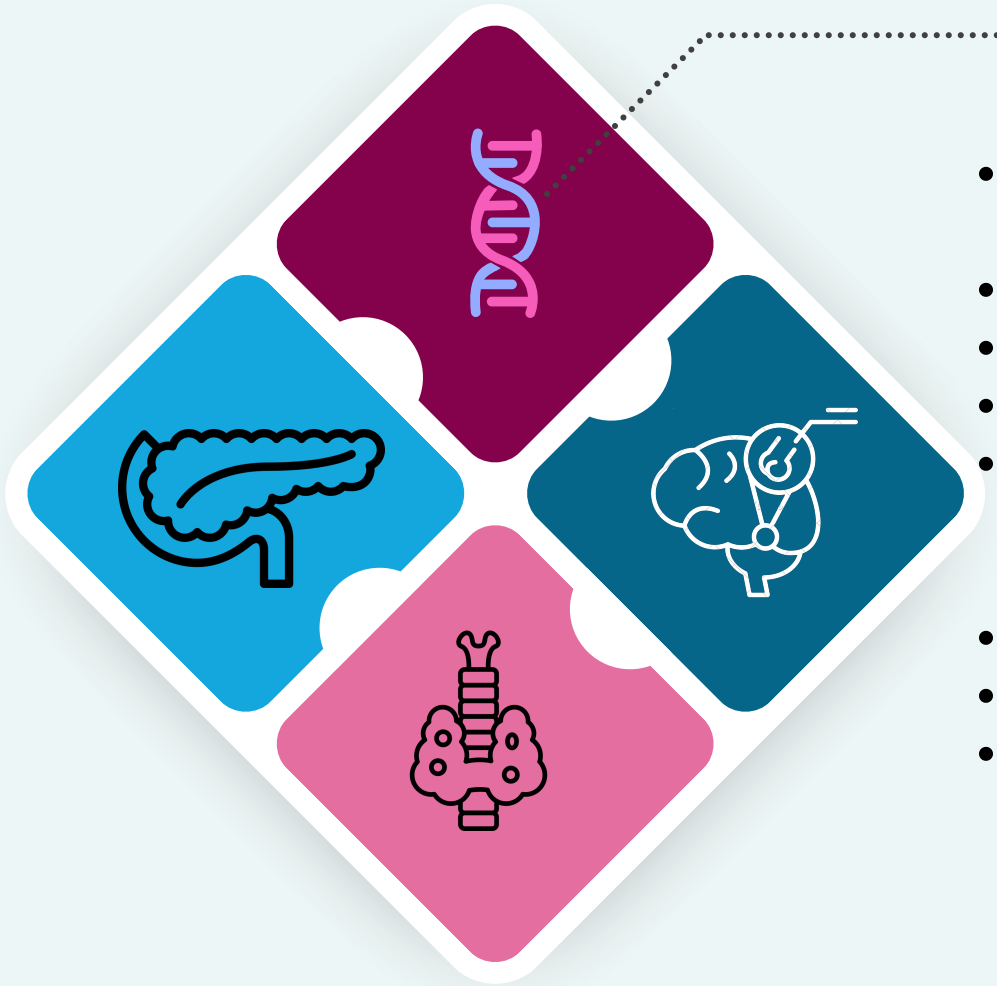

## Indications for Genetic testing / sporadic MEN 1 related tumor

- Parathyroid adenoma < 30 years or multigland disease
- Gastrinoma
- Multiple pancreatic NETs at any age
- ≥ 2 MEN 1 associated tumours
- Thymic NET at any age

**New Considerations 2025 :**

- Solitary pancreatic NET< 40 years
- Non-functioning NET > 1 cm or functioning
- Pituitary adenoma (except microprolactinoma in women) < 30 years

# Biochemical Screening

| | Age to begin biochemische screening | Interval | Recommended screening biochemical test |
|---|---|---|---|
| **Parathyroid tumours** | 10 years | annual | Calcium |
| **Pituitary adenoma** | 10 years | annual | Prolactin, IGF-1 |
| **Duodenopancreatic neuroendocrine tumors** | | | |
| **Non-functioning pancreatic NET** | None | None | None |
| **Gastrinoma** | > 18 years | annual | fasting Gastrin |
| **Insulinoma** | 5 years | annual | None |

**It is explicitly recommended that all MEN patients in all age groups receive care at a specialist centre.**

# Imaging screening

| | Age to begin imaging screening | Interval | Recommended Screening imaging modality |
|---|---|---|---|
| **Parathyroid tumours** | None | None | None |
| **Pituitary adenoma** | 15 years | Every 3-5 years | MRI Pituitary |
| **Adrenal tumours** | 10-15 years | Every 2-3 years | MRI Abdomen |
| **Thoracic NET** | 20-25 years | Every 3-5 years | CT Chest |
| **Duodenopancreatic neuroendocrine tumors** | | | |
| **Non-functioning pancreatic NET** | 10-15 years | Every 2-3 years | MRI Abdomen |
| **Gastrinoma** | None | None | None |
| **Insulinoma** | None | None | None |
| **It is explicitly recommended that all MEN patients in all age groups receive care at a specialist centre.** | | | |

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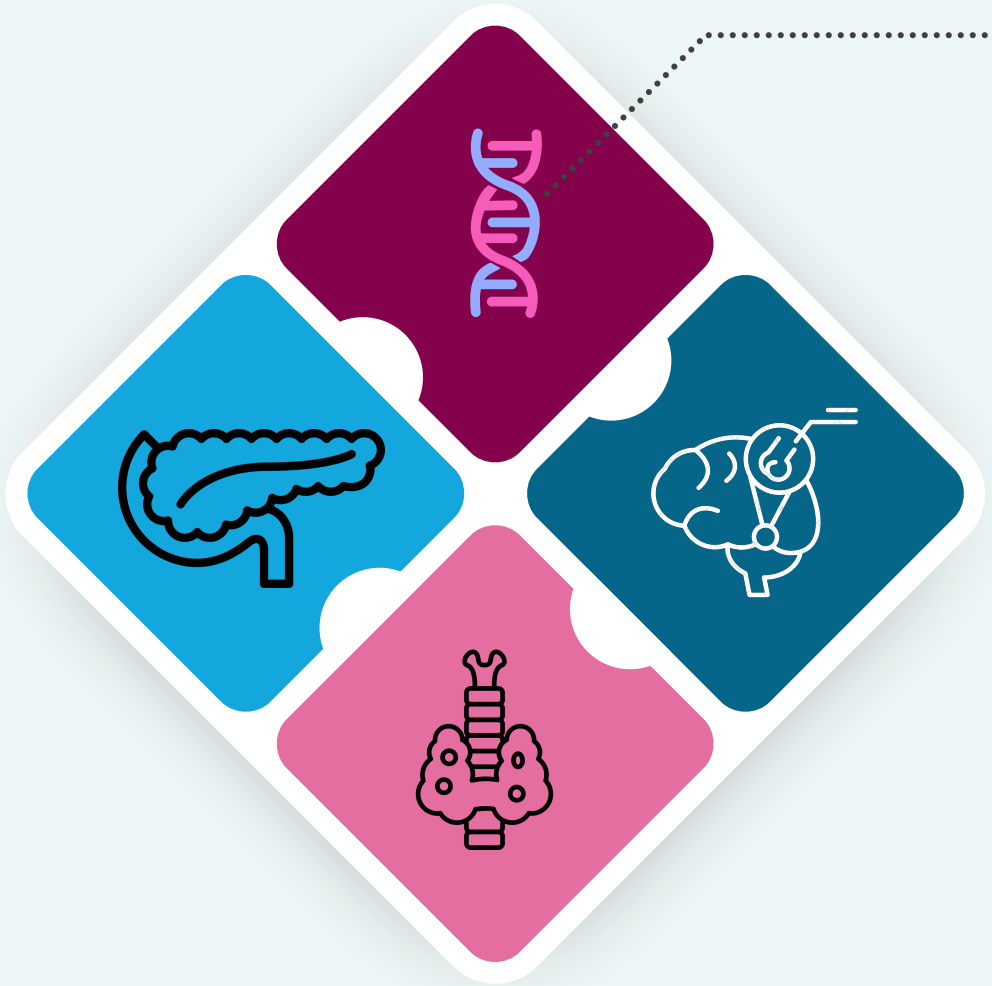

## Recommendations for children

**Clinical evaluation** in consultation with paediatric endocrinologists about 5 years of age:
Education of parents and carers and primary care physicians for awareness of hypoglycaemic symptoms and deviation from normal patterns of growth and puberty at an early age by

- Tracking height and weight percentiles
- Puberal development assessments

**Genetic counselling** to determine age of genetic testing by **shared decision** making with parents and carers, preferably in the first decade of life.

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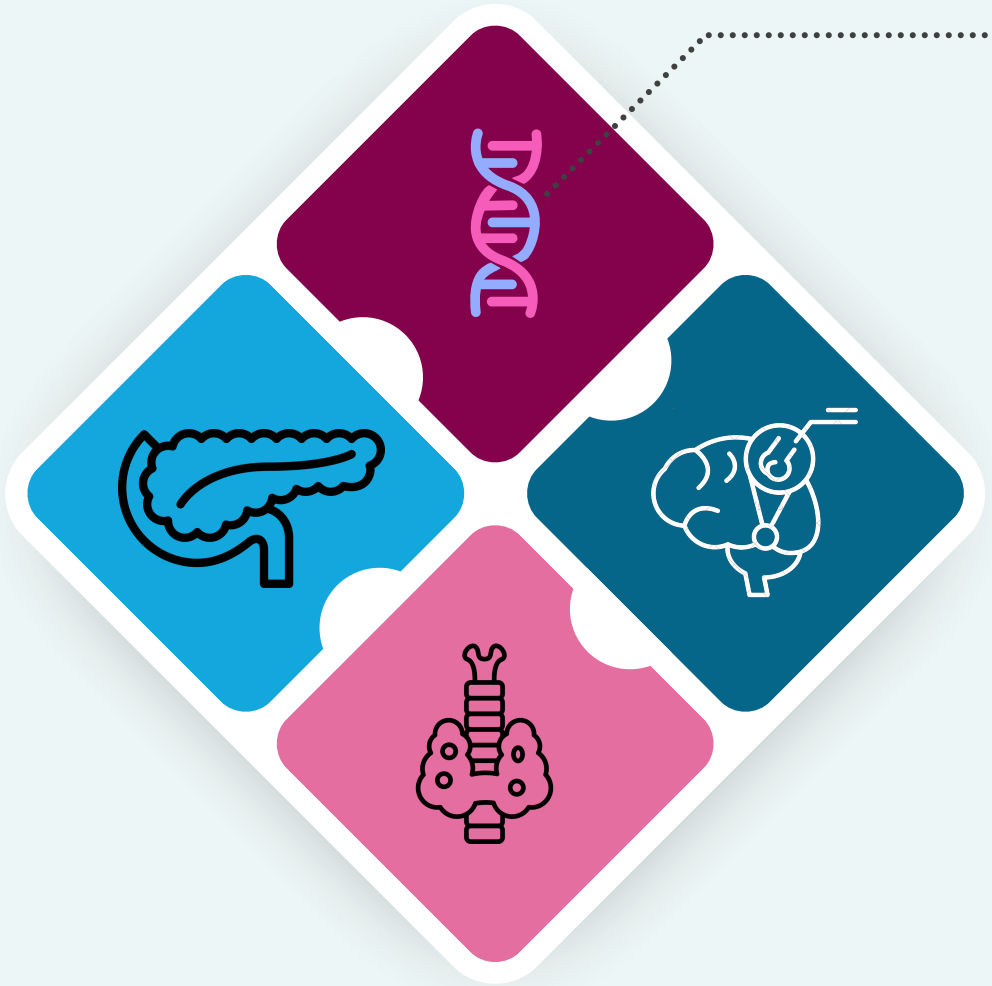

## Recommendations for children

If the MEN 1 mutation is identified or the decision made no to undertake genetic testing in childhood, this screening plan is recommended:

**Biochemistry**: consider initial screening at age 10 years.
**Parathyroid**: Calcium, every 1-3 years
**Pituitary:** Prolactin, IGF1, every 1-3 years
**Pancreas and duodenum**: none

**Imaging:** consider initial screening
**Pituitary:** MRI from age 15, every 3-5 years
**Pancreas and duodenum:** abdominal MRI from ages 10-15 years, every 2-3 years

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## Acknowledgements

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