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ECT – Multiple Endocrine Neoplasia Type 1 and Pancreatic Neuroendocrine Tumours

NIHR BioResource – Rare Diseases study project

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Summary

Pancreatic neuroendocrine tumours (PNETs) may occur as a non-familial (that is, sporadic) isolated endocrinopathy, or as part of a complex hereditary syndrome such as multiple endocrine neoplasia type 1 (MEN1).

MEN1 is characterised by the combined occurrence of PNETs, with tumours of the pituitary, parathyroids, adrenals and NETs of other organs such as the thymus, lungs, bowel and ovary. The majority of patients with MEN1 will have PNETs and somatic *MEN1* mutations are found in >40% of sporadic PNETs.

The aim of this study is to establish a national database of patients with PNET and MEN1, as this will help to:

1. provide important clinical information that will help to inform the epidemiology, natural history, genetics and biology of these diseases;
2. inform on outcomes of investigations and treatments;
3. formulate clinical practice guidelines that will improve patient care; and
4. further our understanding of the mechanisms underlying these disorders.

The national database will be of benefit to doctors and researchers investigating the epidemiology, genetics and cellular mechanisms of these diseases, as well as identifying suitable patients for clinical trials, aimed at assessing novel therapeutics. Patients and patient support groups will also benefit from improved care that will result from the clinical guidelines that will likely be generated by database.

Patient group: AMEND - The Association for Multiple Endocrine Neoplasia Disorders https://www.amend.org.uk/

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Recruitment Criteria

Inclusion

A patient of any age or sex with pancreatic neuroendocrine tumours (PNETs) or multiple endocrine neoplasia type 1 (MEN1).

A relative of any age or sex of an affected patient.

Exclusion

Exclusion / withdrawal criteria include ineligibility (either arising at recruitment, during the study or retrospectively having been overlooked at screening).