Audit of Patients with Multiple Endocrine Neoplasia Type 1 (MEN1): Screening of Pancreatic Neuroendocrine Tumours (pNETs), Parathyroid Tumours and Pituitary Adenomas

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INTRODUCTION

Patients with MEN1 have increased morbidity and mortality compared to those patients with sporadic NETs.
No genotype-phenotype correlation is described and age-related clinical penetrance surpasses 50% and 90% by 20 and 40 years, respectively.
The aim of the audit was to compare the screening programme for MEN1 patients with MEN1 clinical guidelines.

METHODS

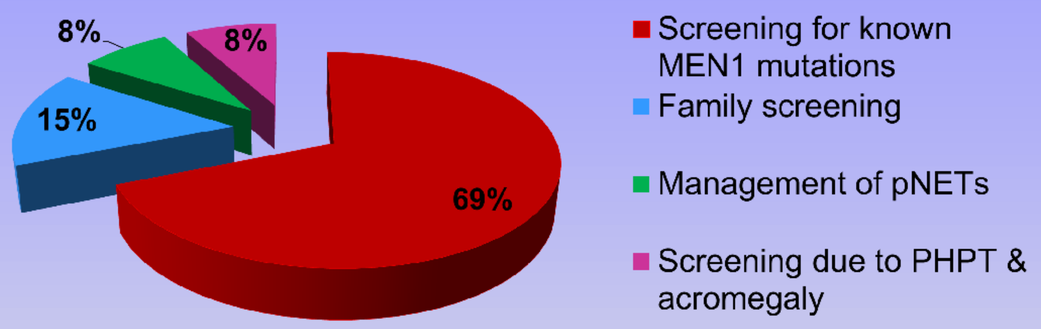
Case notes of MEN1 patients attending a tertiary NET-multidisciplinary team (MDT) in Ireland were reviewed.

All patients attending the NET-MDT have gastrointestinal hormone, parathyroid, pituitary profiles, endoscopic and imaging studies according to guidelines.

RESULTS

Patients with MEN1
•13 patients (11 kindreds), 100% Caucasians, mean age 43 years (range 28-67): 69% male, 31% female
•85% (n=11) had confirmed MEN1 mutations and two had clinical or familial MEN1

Reasons for Patients referred to NET-MDT



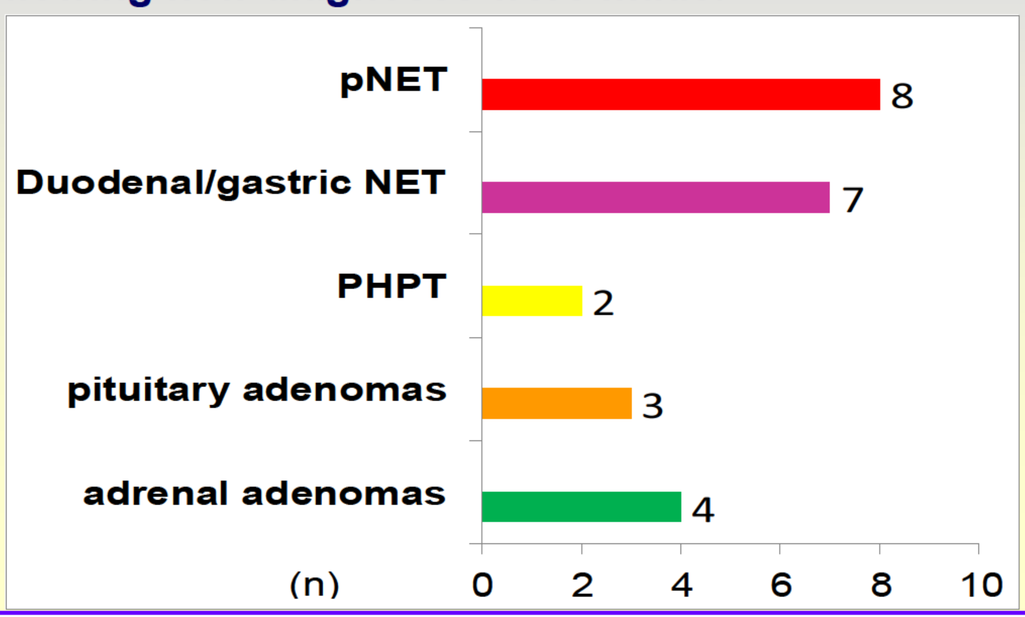
Prior to NET-MDT assessment,

- 46% did not have endoscopic/radiology studies to screen for pNETs
- none of the patients had CT/MRI thorax to screen for thymic/bronchial NETs

Age at screening

- age referred to NET-MDT: 41.5±12.2 years.
- age at endoscopic/radiological screening for NETs (prior to/at NET-MDT): 37.1±14.3 years
- age at diagnosis of NETs: 34.9±14.3 years

On screening of NET-MDT, the following new diagnoses were made:



CONCLUSIONS

Endoscopic/radiological screening of NETs occurred at later age than recommended by current guidelines.
Surveillance methods were also largely at variance with guidelines.
Referral to a dedicated MDT has identified a significant number of previously unrecognised neuroendocrine pathologies.

References

Guidelines for diagnosis and therapy of MEN type 1 and type 2. Brandi et al, J Clin Endocrinol Metab. 2001;86(12):5658-71
Clinical Practice Guidelines for Multiple Endocrine Neoplasia Type 1 (MEN1) 2012, Thakker et al, J Clin Endocrinol Metab, 2012;97:2990–3011

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