# Patients with multiple endocrine neoplasia type 1 (MEN1) have late progression and long survival despite the presence of disseminated disease: the experience of a referral center in Greece

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## BACKGROUND

**Multiple endocrine neoplasia type 1 (MEN1) is a genetic disorder involving mainly parathyroid tumors, pancreatic neuroendocrine neoplasms (pNENs) and pituitary tumors**

## AIMS

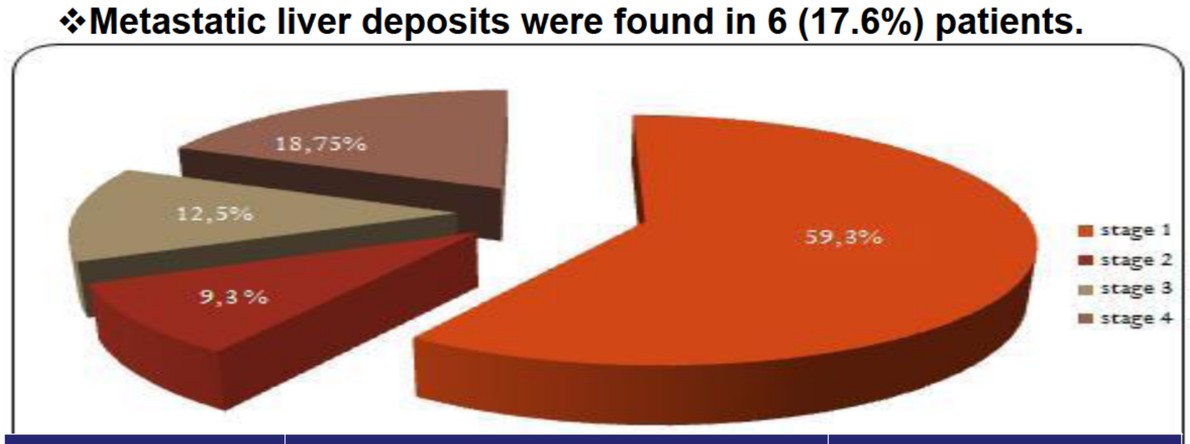
**Registration of demographic, clinical, imaging, pathological features, therapeutic options, response to treatment, overall survival of patients with MEN1.**

## METHODS

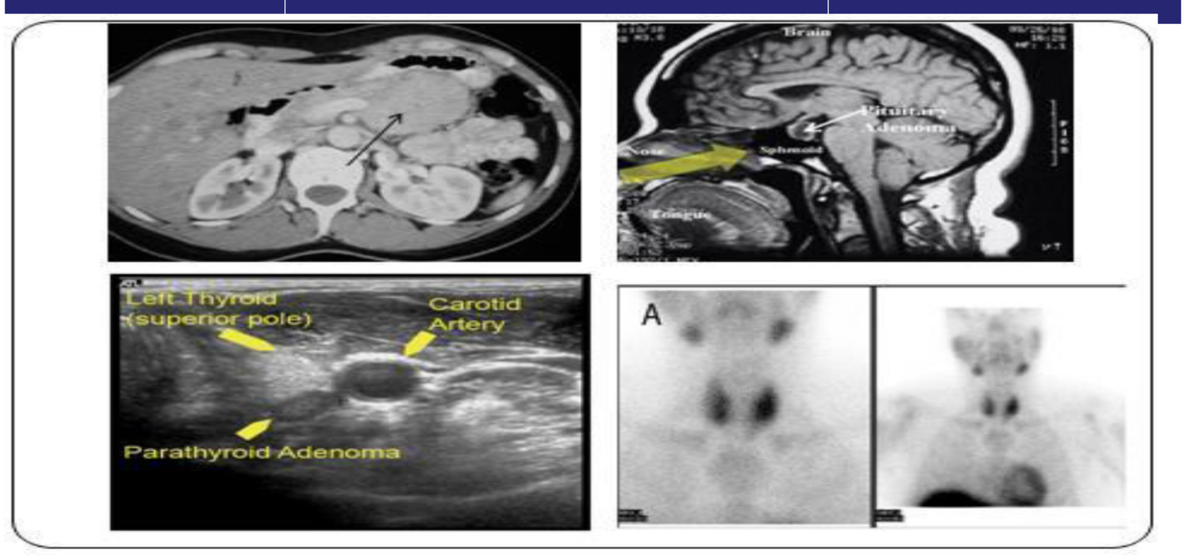
- ***35 patients with* MEN1 *from our data-base registered during the period 2004-2014 have been retrospectively studied***
- ***TNM classification system has been used for staging***
- ***Proliferation index Ki-67 has been used for grading***
- ***Registered: primary site, presence of secretory/ functional syndrome, metastatic deposits, presence of familiar syndrome***
- ***Therapeutic management and outcome were registered***

## RESULTS

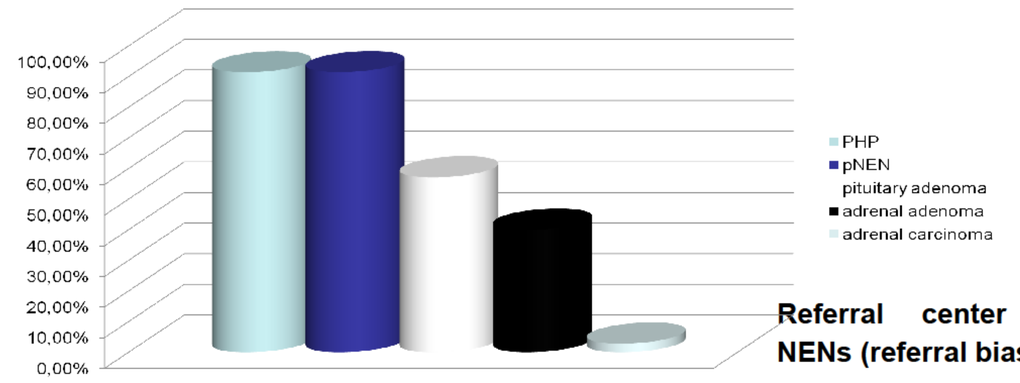
- **24 (68.6%) patients were the index cases.**
- **19 (54.3%) had a positive gene mutation while 5 (14.3%) did not have the mutated gene.**
- **11 (32.4%) patients had a functional syndrome.**
- **Metastatic liver deposits were found in 6 (17.6%) patients.**




| | | |
|---|---|---|
| Age | | 39 (15-64) years |
| Gender | Males<br>Females | 20<br>15 |
| Syndrome components | Primary hyperparathyroidism | 32 |
| | Pituitary Adenoma | 20 |
| | NFPA | 11 |
| | PRLoma | 8 |
| | GH | 1 |
| | Pancreatic Neuroendocrine Neoplasm | 32 |
| | Adrenal Adenoma | 14 |
| | Aarcinoma | 1 |
| | Fibromata | 3 |




## RESULTS




**Referral center for NENs (referral bias).**

**pNENs**

| | | |
|---|---|---|
| Grade | 1<br>2<br>unknown | 11<br>9<br>12 |
| Non fumctioning<br>Gastrinoma<br>Insulinoma | | 23<br>7<br>2 |
| Stage | 1<br>2<br>3<br>4 | 19<br>3<br>4<br>6 |
| Surgical removal | | 14 |

| Line of treatment | 1st | 2nd | 3rd | 4th | 5th | 6th |
|---|---|---|---|---|---|---|
| Surgery | 14 | 4 | 2 | | | |
| Somatostatin receptors analogs | 6 | 8 | 3 | 3 | 3 | |
| Chemotherapy | 2 | 3 | 1 | 1 | 1 | 1 |
| Molecular targeted therapy | 1 | 2 | 3 | 2 | | |
| Peptide receptor radionuclide therapy | 1 | 1 | 1 | 1 | | |
| Chemoembolization | 1 | | 1 | | 1 | |
| Follow up only | 21 | | | | | |

**Molecular targeted therapy: everolimus, sunitinib.**
**Chemoembolization: Transarterial Chemoembolization (TACE) or Transarterial Embolisation (TAE).**

**27 patients are alive with mean follow-up time 94,64 months (range 10-316.33) since diagnosis**

**3 (9,67%) patients died of their disease out of the 31 under current follow-up with a mean survival time 121 months (range 62.3-190) since diagnosis:2 with grade 1 pNEN and 1 grade 2 pNEN-1 patients died during surgery (pulmonary embolism)**

| Patients | Gender | Functional syndrome | Initial therapy | Stage | Grade | Liver mets | Bone mets | Octreoscan | PHP | Pituitary adenoma | Adrenal adenoma |
|---|---|---|---|---|---|---|---|---|---|---|---|
| 1 | A | - | Follow up | 2 | 2 | - | - | + | + | + | bilateral |
| 2 | A | + | Follow up | 3 | Unknown | - | - | + | + | + | - |
| 3 | A | _ | SSA | 4 | 2 | + | - | + | + | + | unilateral |

## Conclusion

**The present registry implies that the majority of the patients with MEN1 have late progression and long survival despite the presence of disseminated disease, confirming the necessity of specific therapeutic and diagnostic options following the guidelines as well as their management from referral centers under multidisciplinary teams.**

**References**Thakker et al. J Clin Endocrinol Metab. 2012 Sep;97(9):2990-3011

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