

The New Zealand Cancer Registry is not a complete record of neuroendocrine malignancy.

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Background

Neuroendocrine tumours (NETs) are a heterogeneous group of malignancies arising from cells of the diffuse neuroendocrine system. They exhibit a clinical course ranging from indolent to highly aggressive. Histological classification of NETs has been subject to multiple modifications over recent decades¹. NET classification prior to 2000 also assumed that many NETs were benign but now all are considered malignant. As the New Zealand Cancer Registry (NZCR) only mandates reporting of malignant tumours, this improved understanding of NETs has likely led to historical NET diagnoses being unrecorded on the NZCR.

The NETwork! Study is a national programme that aims to improve clinical outcomes for patients with NETs and includes an epidemiological description to guide future service provision. This preliminary study describes early data from the NETwork! Project, focussing on the NZCR-reported NET incidence in NZ.

Aims

- To identify NET cases in the Auckland District Health Board (ADHB) area;
- To investigate the incidence and epidemiology of NETs; and
- To assess the extent to which the NZCR can be considered a comprehensive record of NETs.

Methods

We conducted a retrospective review of all NET diagnoses from 2002 to 2011 for patients living in the ADHB catchment. Hypothesising that the NZCR would not be a complete record of NETs, we reviewed the Auckland City Hospital LabPlus Delphic Anatomical Pathology database of reports as well as the NZCR.

Cases identified by the two search strategies were processed to yield a combined list with indications of whether each NET had been recorded on the NZCR and/or the pathology database. For NETs not registered with the NZCR, a review of clinical records was conducted to retrieve data for analysis such as sex, ethnicity, date of birth, date of diagnosis, primary site of tumour, tumour histology and date of death.

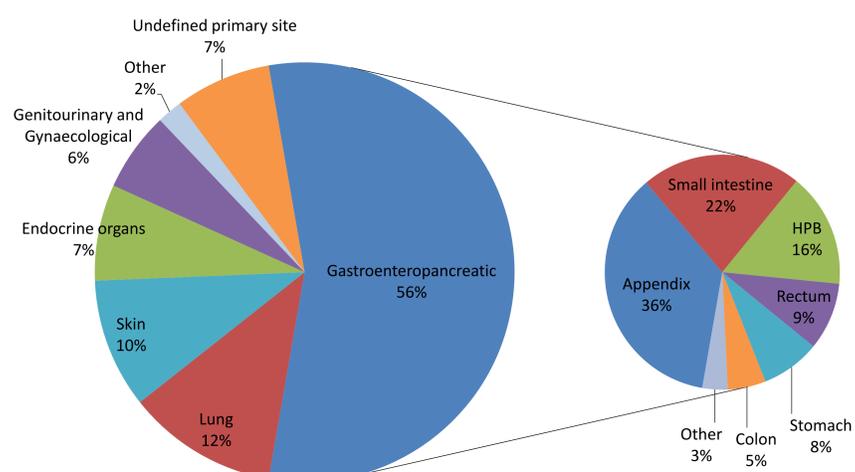
Results

A total of 310 NET cases were identified. The NZCR identified 209 (67%) cases. An additional 101 (33%) were identified in the ADHB pathology database and had not been recorded on the NZCR.

There were 167 (54%) female patients with NETs and 143 (46%) male patients. Median age at diagnosis was 61 (mean age at diagnosis 58; standard deviation 20). Distribution of NETs by ethnicity was 67% European, 12% Māori, 9% Pacific peoples, 7% Asian, 1% Middle Eastern and 0.3% African. The remaining 3% of patients did not state ethnicity.

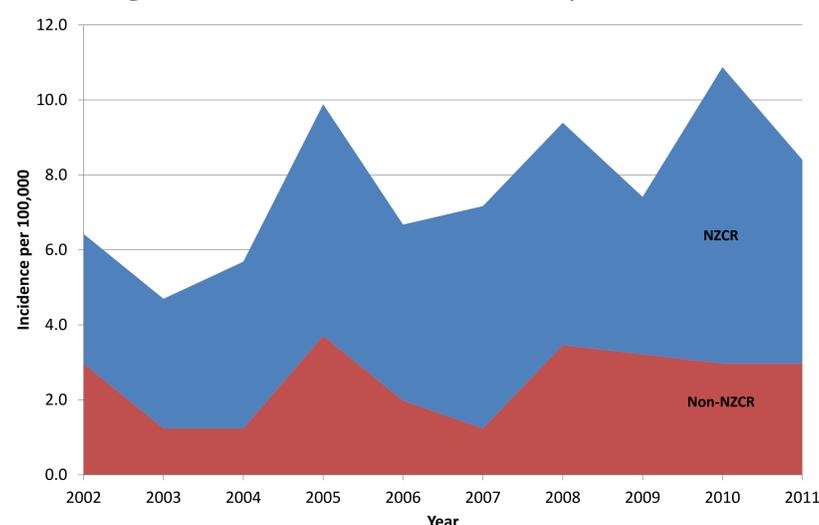
In assessing case-reporting to the NZCR by patient ethnicity we found that for European patients 150/207 (72%) of NET cases were reported. However, for Māori patients this was 17/38 (45%) and for Pacific patients 17/29 (46%).

Figure 1. NETs by primary site (n=310) with breakdown of gastroenteropancreatic NETs (n=172)



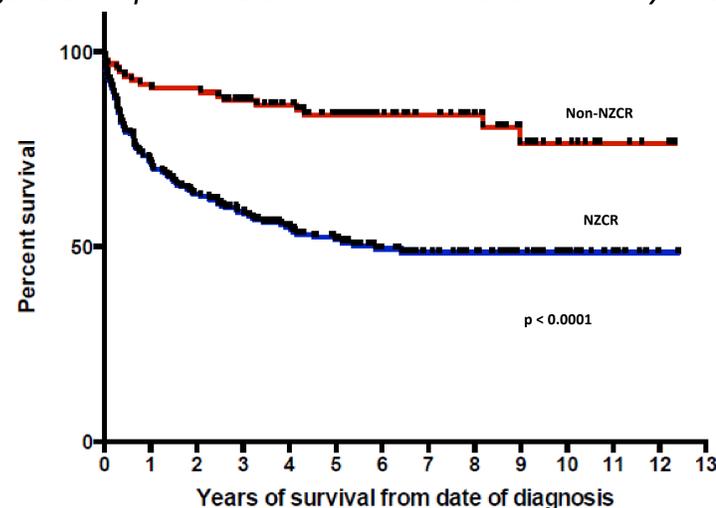
The majority of NETs were from gastroenteropancreatic primary sites. Lung NETs were large cell and carcinoid tumours. Small cell lung carcinomas were excluded. The endocrine organs group consisted of pheochromocytomas, paragangliomas and medullary thyroid carcinomas. Liver, pancreatic, gall bladder and other biliary tract NETs were grouped as hepatopancreaticobiliary (HPB) NETs.

Figure 2. Annual NET incidence by database



The red area of the graph indicates incidence as calculated from NETs identified through the pathology database search (i.e. NETs that were not registered on the NZCR). The blue area stacked above indicates the additional incidence from NZCR NETs. Total NET incidence increased from 2002 (6.4/100,000) to 2011 (8.4/100,000). Over the entire ten-year period the average annual incidence was 7.7 per 100,000.

Figure 3. Kaplan-Meier survival curve: survival by database



NZCR NET cases had a median survival of 5.9 years, significantly shorter than that of the non-NZCR cases ($p < 0.0001$). Non-NZCR NET survival was undefined as survival exceeded 50% at time of analysis. This suggests the unreported NETs are the more indolent tumours that might not have required reporting under the previous WHO classification.

Conclusions

The NZCR is a vital source of cancer data but is not a complete record of NETs. In this cohort, NZCR data underestimates NET incidence by 33%. An accurate estimate of NET epidemiology will require interrogation of multiple data sources including pathology databases. NZCR users in the health policy sector should be aware of these limitations when designing NET service provision.

This single-DHB study has also provided an estimate of NET incidence in a NZ population. In ADHB, NET incidence is increasing, a finding in line with overseas studies^{2,3}. Median survival differs significantly between NZCR and non-NZCR NETs. This is likely due to the relatively recent recognition of NETs as malignant tumours. Small, well-differentiated and incidental NETs were previously considered benign and thus, in line with our understanding at the time, would not have been reported to the NZCR.

Our study is currently expanding to other DHBs and will use additional databases that will modify final epidemiological characterisation. Preliminary epidemiological findings also require further investigation, for instance, under-reporting associated with ethnicity. This project is a part of the NET Epidemiology in NZ study that will ultimately inform the design of a new proposed multicentre multidisciplinary model of care for NZ patients with NETs.

References

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