PHAEOCHROMOCYTOMAS AND PARAGANGLIOMAS -
A 20 YEAR EXPERIENCE FROM THE ROYAL SUSSEX COUNTY HOSPITAL

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Aim

To investigate our management of patients diagnosed with Phaeochromocytoma and Paraganglioma over the last 20years and compliance with the following guidelines:

- The NANETS Consensus Guideline for the Diagnosis and Management of Neuroendocrine Tumors: Phaeochromocytoma, Paraganglioma & Medullary Thyroid Cancer- 2010[1]
- Phaeochromocytoma: Recommendations for clinical practice from the First International Symposium-2005, at Bethesda, MD, USA [2]

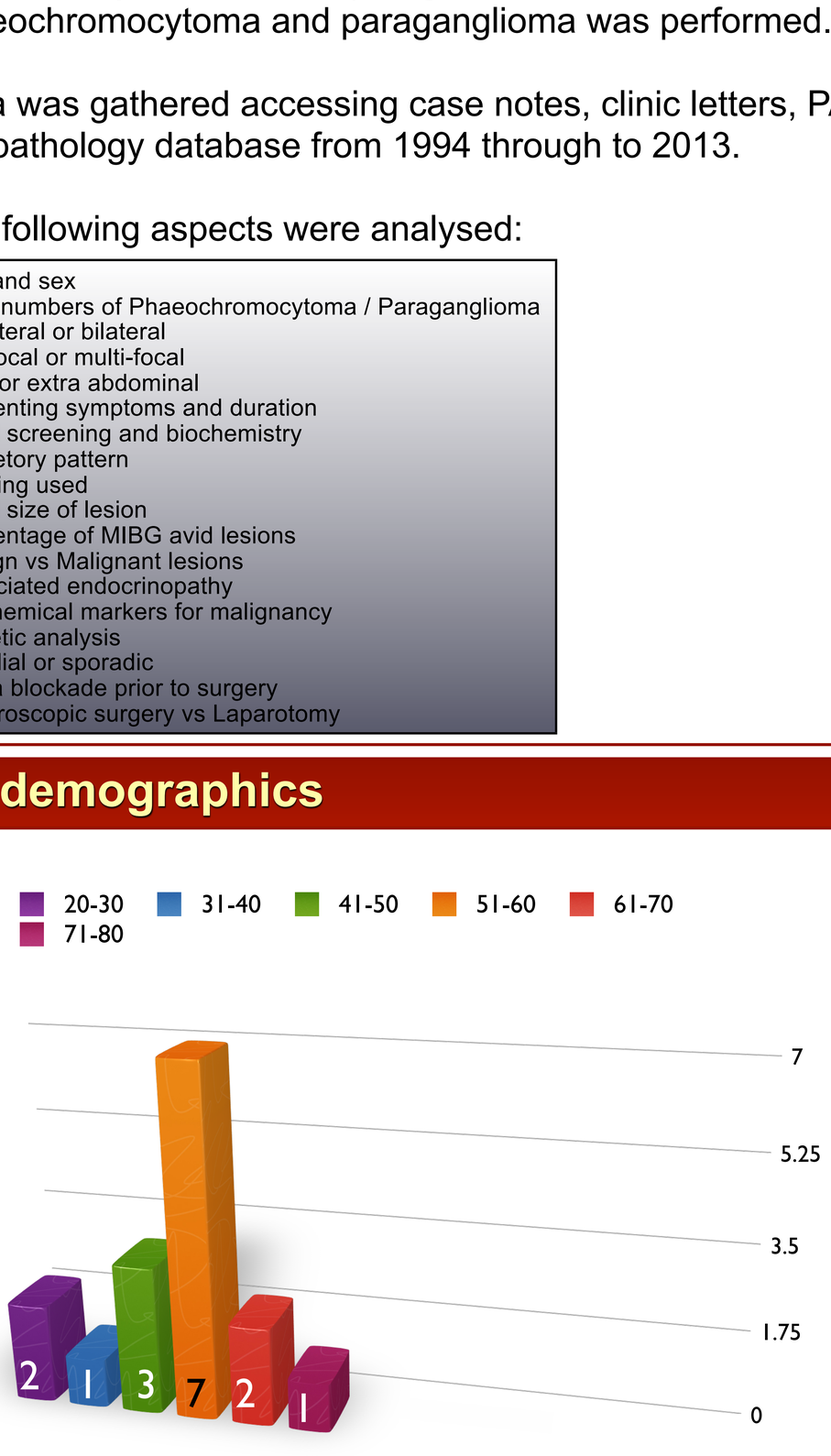
Introduction

- Phaeochromocytomas and paragangliomas account for about 0.1% of cases of persistent hypertension.[1]
- Only 50% of these are being diagnosed as symptoms are often paroxysmal.[1]
- The initial screening tests, imaging, genetic analysis and modes of treatment have evolved significantly over the last two decades.
- Nevertheless, a standardised approach is essential for early diagnosis and decision regarding appropriate treatment for this diverse condition.

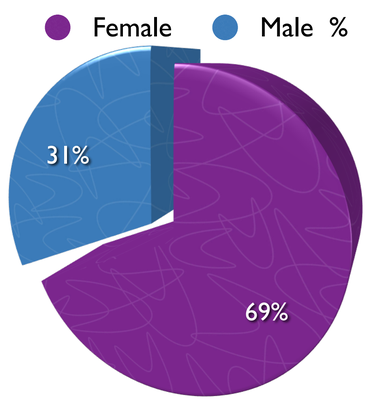
Methods and Materials

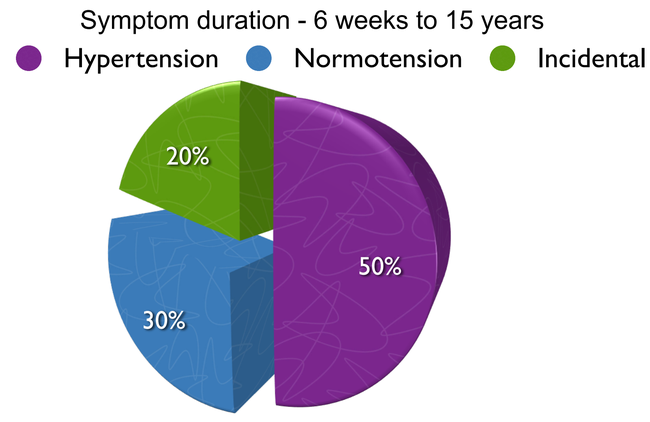
- A retrospective data analysis of 16 random patients (Age 24-71 years, mean51) diagnosed and treated for phaeochromocytoma and paraganglioma was performed.
- Data was gathered accessing case notes, clinic letters, PACS and our pathology database from 1994 through to 2013.
- The following aspects were analysed:
  - Age and sex
  - Total numbers of Phaeochromocytoma / Paraganglioma
  - Unilateral or bilateral
  - Uni-focal or multi-focal
  - Intra or extra abdominal
  - Presenting symptoms and duration
  - Initial screening and biochemistry
  - Secretory pattern
  - Imaging used
  - Initial size of lesion
  - Percentage of MIBG avid lesions
  - Benign vs Malignant lesions
  - Associated endocrinopathy
  - Biochemical markers for malignancy
  - Genetic analysis
  - Familial or sporadic
  - Alpha blockade prior to surgery
  - Laparoscopic surgery vs Laparotomy

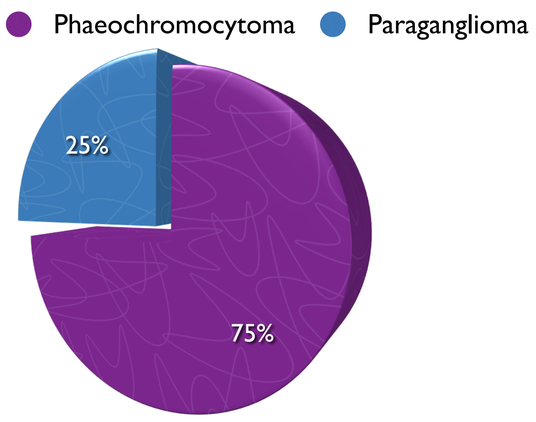
Age demographics



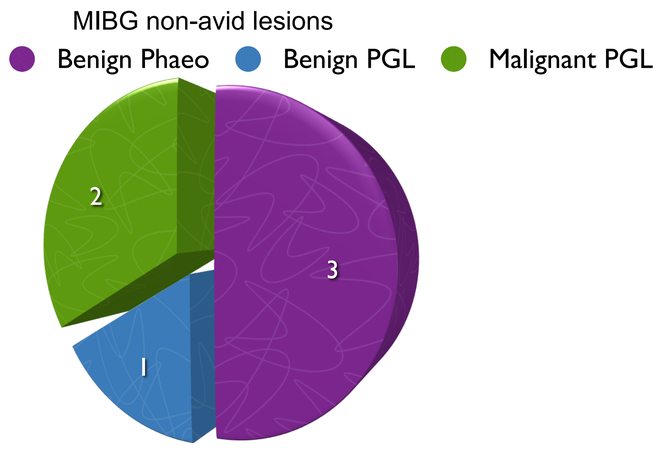
Results







Phaeochromocytoma (75%)	
Unilateral	100%
Right sided	58%
Malignancy	8%
Initial size on CT	0.5-10cm (mean 5cm)
Secretory pattern	Adrenaline - 67% Adrenaline +Noradrenaline - 58% Dopamine - 16%
Occurrence	Familial - 8% Sporadic - 92%
α-blockade	80%
Surgery	Laparoscopy - 67% Laparotomy - 33%



Paraganglioma (25%)	
Location	50% intra abdominal 50% extra abdominal
Malignancy	75%
Secretory pattern	Noradrenaline - 100% Dopamine - 75%

- All but 1 patient under 50 years of age were referred for mutation analysis as per guideline.[2]
- Mutation analysis was negative for SDHB, SDHC, SDHD, VHL and RET.
- 1 patient was diagnosed with neurofibromatosis -1.
- 80% of patients received α-blockade with Phenoxybenzamine during the first consultation and the rest treated with other agents subsequently.
- All patients with Phaeochromocytoma were able to discontinue their α-blockade post surgery.
- Urine catecholamines was the biochemical screening tool used for all patients (100%). Urine metanephrines (13%) VMA (6%) was also used. Plasma metanephrines was not used in any of them.
- Urine dopamine when elevated, proved to be a highly sensitive marker to predict malignancy in our cohort of patients. This has been confirmed previously in several reports.[5]
- CT / MRI was the initial imaging modality followed by MIBG[2] in 63% of patients. The remaining had MIBG as first line.
- FDG-PET[2] was used in two patients (13%)(1 benign Phaeo) and 1 malignant PGL as the lesions were MIBG non-avid.
- Histology was reported by various pathologists and features of local invasion did not always correlate with malignancy.[2,7]

Conclusions

- Our study is a snap shot that reflects the general trend of management of these tumours in various centres in the UK.
- There is an emerging evidence of superiority of metanephrines,[3,4] especially plasma over urine as a screening tool and there is a growing trend not to rely solely on urine catecholamines when the clinical suspicion is high.
- Predominantly dopamine secreting adrenal tumours could be MIBG non-avid and other functional imaging modalities should be considered; especially to address the increased risk of malignancy.[5,6]
- The evolving genetic spectrum of mutations for screening including TMEM127, MAX, Fumarate hydratase and various others propose an additional challenge for the clinician and the geneticist.
- A standardised histopathology report with the inclusion of PASS[7] or other similar scoring system might speak the same language in a multi-disciplinary setting to decide an appropriate management plan.

A re-audit with the above considerations will be performed in the next 5 years.

References

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