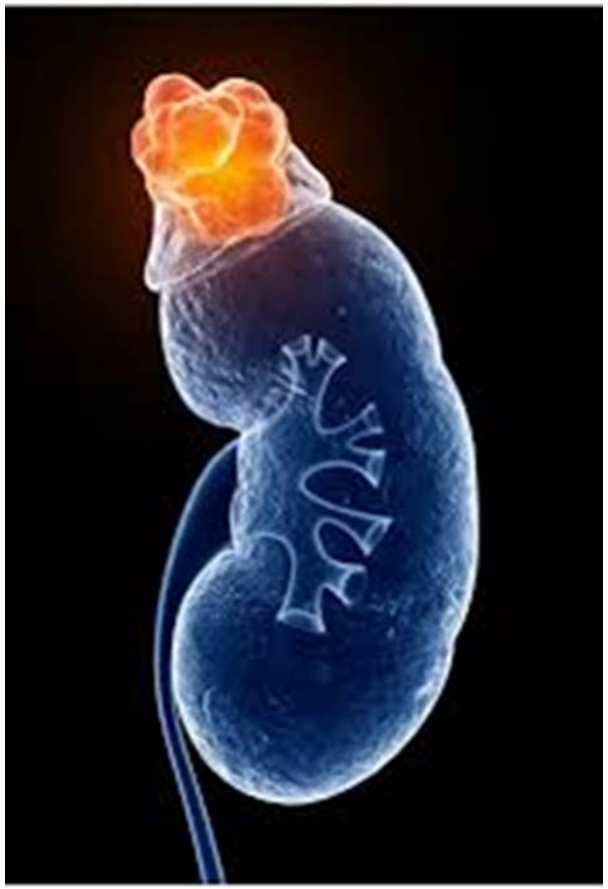
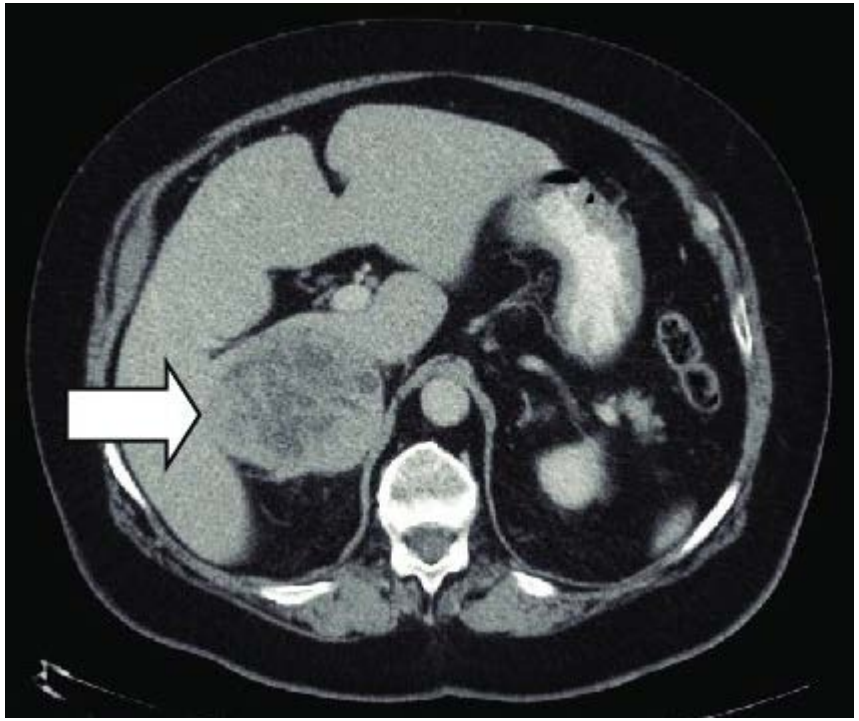


Phaeochromocytoma in Pregnancy

Dr. L. Cairncross Consultant and Head: Endocrine and Breast Surgical Unit, Division of General Surgery, UCT

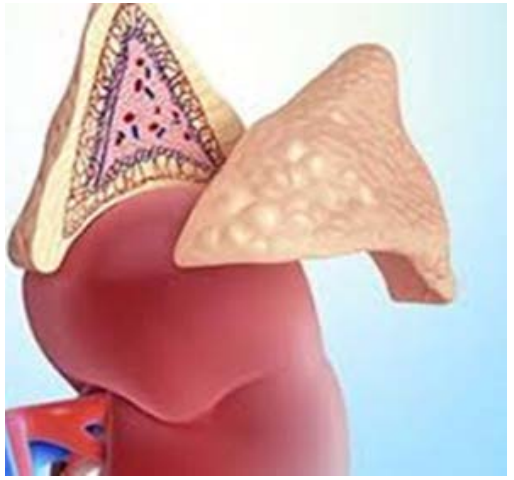
A Patient's Story….

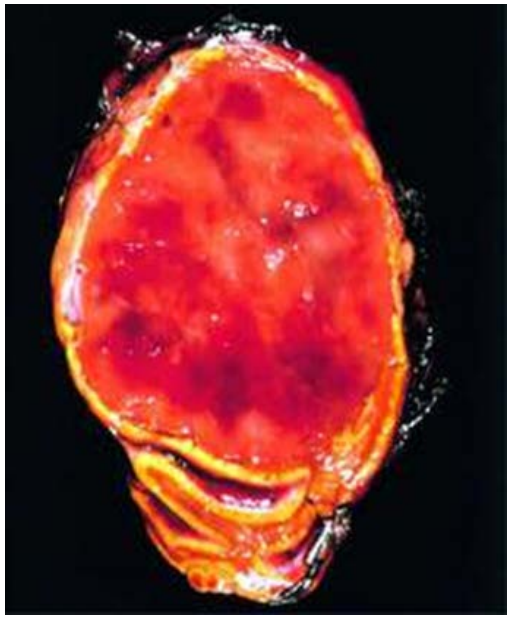
- 24 yr old G3 P1 33/40 "pre eclampsia" – BP 230/170
- D3 foetal distress – C/S 1.4kg baby, died within 48h
- D7 postpartum hypertensive crisis - ICU
- Hx: G1 at 17, "pre eclampsia" and stillbirth, G2 miscarriage
- No oedema, no proteinurea
- Urine catecholamines ⬆ and CT – 5 cm R adrenal mass
- Open adrenalectomy and subsequent normal pregnancy.



Definitions:

- Phaeochromocystomas and functional paragangliomas - rare (0.2% hypertensives)
- Catecholamine secreting tumours arising from chromaffin tissue
- 80 – 90% adrenal medulla (phaeochromocytomas)
- Anywhere along the sympathetic chain (paragangliomas)
- Paragangliomas – noradrenaline, more likely to be malignant, associated genetic disorders
- Phaeochromoytoma here refers to both, though paragangliomas in pregnancy – better prognosis.





Presentation in Pregnancy:

- 5 and 10% of pregnant women are hypertensive
- Pre-existing hypertension or pregnancy associated hypertension
- Phaeo's 2-7/100 000 pregnancies, most midwives and obstetricians will not see this condition
- Pre delivery diagnosis reduces mortality

PREECLAMPSIA
foundation

Clinical Presentation Phaeochromocytoma:

- Pregnancy same classic triad presentation:
  - Diaphoresis (abnormal whole-body sweating)
  - Palpitations
  - Headache

- Hypertensive crisis:
  - acute pulmonary oedema
  - malignant arrhythmias
  - myocardial ischemia or infarction
  - aortic dissection
  - cardiac failure
  - haemodynamic collapse

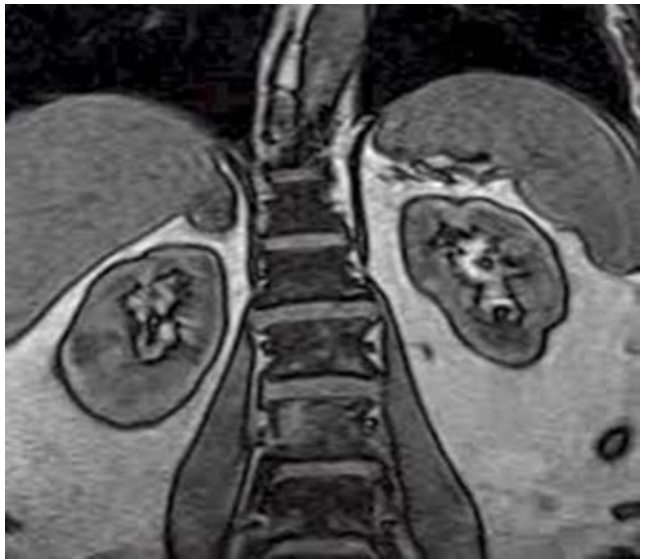
Phaeochromocytoma vs Pre-Eclampsia:

Phaeochromocytoma:

- May present before 20 weeks
- Paroxysmal hypertension
- No oedema and usually no proteinuria.
- Episodes of postural hypotension (up to 50%)
- Gestational diabetes and cardiomyopathy
- Increasing severity advancing gestational age
  - pressure of the growing uterus on the tumour
  - foetal movements
  - contractions

Diagnosis in Pregnancy:

- Catecholamine release not affected by pregnancy
- 24 hour urine metanephrines and noremetanephrines reliable
- As with non-pregnant patients, 3 – 4 x normal is diagnostic
- MRI are the imaging modality of choice
- Nuclear medicine studies such as MIBG/Dota-scans contraindicated
- Up to 30% phaeo pts inherited genetic disorder
- Pregnant patients - younger cohort of patients - genetic testing
  - MEN 2a and b
  - Von Hippel Lindau Syndrome (Tumour Suppressor gene Chromosome 3)
  - Neurofibromatosis
  - Succinate dehydrogenase genes
- PTH and calcitonin levels



Management of Phaeo in Pregnancy:

Depends on timing of diagnosis:

1.	Early antenatal	<24/40weeks	(35%)
2.	Late antenatal	> 24/40	(40%)
3.	Intrapartum	labour/CS	(25%)

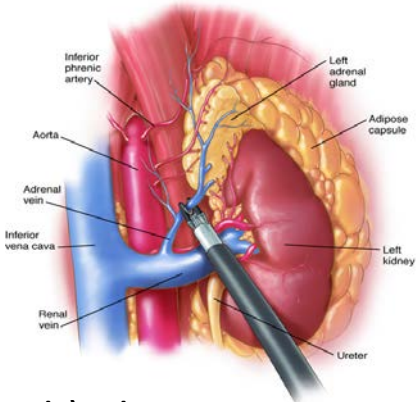
Key Aspects of Management:

a) Medical management

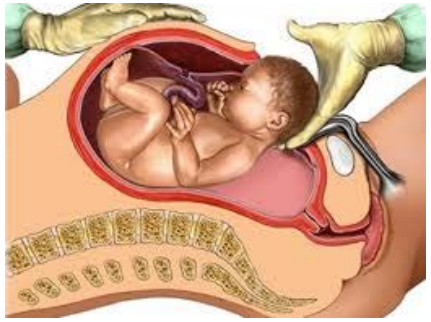
b) Surgery for phaeochromocytoma

c) Timing and mode of delivery of baby

a) Medical Management



b) Phaeo Excision



c) Delivery of Baby

a) Medical Management:

- Alpha blockade – Doxazosin (OR Phenoxybenzamine)

- Intense vasoconstriction – intravascular volume depletion
  - Replace fluids and high salt diet
- No intrinsic vascular placental autoregulation
- Phaeo episodes affect foetus through intense vasoconstriction
  - hypoperfusion, hypoxia and placental abruption.
- Alpha blockade NB but postural hypotension is dangerous
- Targets usually BP <130/80 sitting, standing systolic > 90
- Pregnancy BP targets higher though values unknown

Delicate balance – alpha blockade and hypotension

Other drugs….

<u>B Blockers not routine:</u>

- Persistent tachycardia after adequate alpha blockade and rehydration.
- In pregnancy, B-blockers risk of IUGR
- Calcium channel blockers possible alternative

<u>Caution with other drugs:</u>

- Corticosteroids
- Opiates
- Metaclopramide
- Thiopental
- Ketamine
- Ephidrine

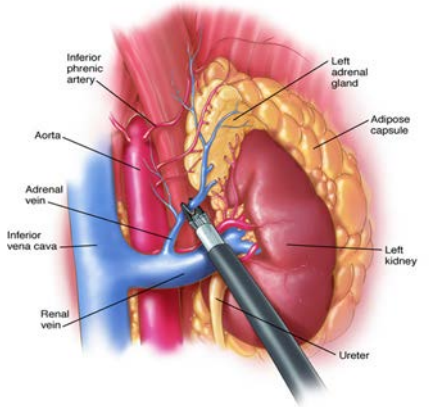
May induce hypertensive crisis

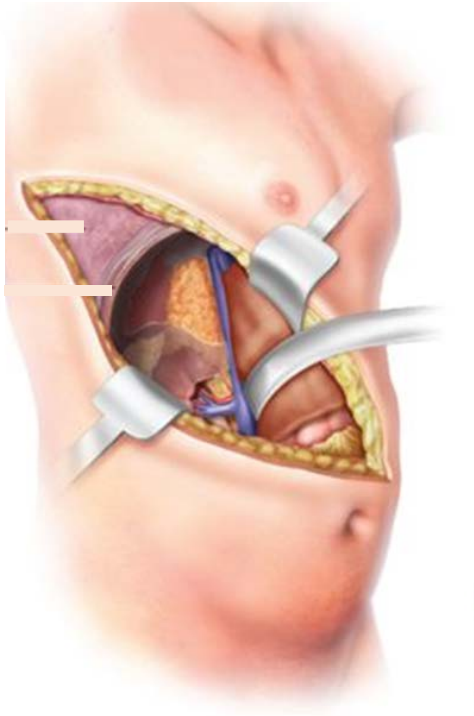
b)Phaeochromocytoma Resection:

Anaesthetic Team Critical!!

Surgical excision is definitive treatment

1. Early antenatal DXN : < 24/40
   - Alpha block and volume replace for 10 – 14 days
   - Surgery second trimester
   - Laparoscopy recommended - only done 50% case reports
   - Individualise: tumour size, size of uterus, experience of team
   - Transperitoneal approach not retroperitoneal
   - Caution left phaeo – right lateral position – IVC compression





2. Late Antenatal DXN: >24 weeks
   - Medical management until delivery
   - After delivery, resect immediately (open or laparoscopic) OR
   - Resect at 3 days or 2-6 weeks

3. Intrapartum DXN: during labour or C/S
   - Very high risk with HPT crisis
   - Deliver baby and stabilise mother
   - Plan resection later

c) Timing and mode of delivery - baby:

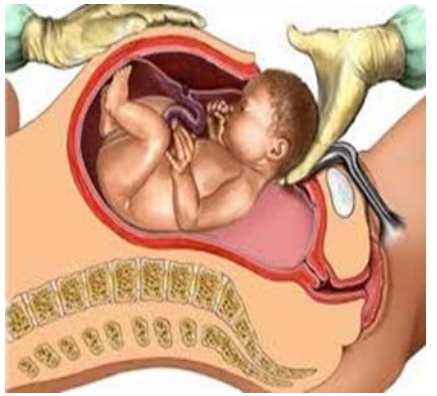
1. Early antenatal DXN <24/40:
   - At term after phaeochromocytoma resection
   - Careful monitoring high risk pregnancy
   - Obstetric considerations for timing and NVD vs C/S
   - Remember possibility of synchronous or metastatic tumours

2. Late antenatal DXN >24/40:
   - Baby delivered with phaeo in situ close as possible to term
   - Monitor foetal growth/well being re timing
   - Caesarian section is generally recommended
   - More controlled environment
   - BUT also risk of anaesthesia, drug administration, haemorrhage – some case reports of NVD in mulitiparous women

3. Intrapartum DXN:
   - Emergency situation, dangerous, with high mortality
   - Deliver baby and stabilise mother – both in ICU
   - Planned resection later

Normal Vaginal Delivery

VS



Caesarian Section

Outcomes:

- 2013 review BJS 77 case reports 2000 - 2012
- Maternal mortality 8%, foetal mortality 17%
- 35% diagnosed early
  - Majority tumour resection second trimester (1/2 laparoscopic, 1/2open)
  - 1/3 after delivery
- 40% diagnosed late in pregnancy
  - 1/2 concurrent phaeo resection with delivery and 1/2 post partum
- Most women delivered by C/S
- 25% Intrapartum DXN – baby delivered first then phaeo resection at 2-8 weeks
- High risk group, only in 12/21 cases did both mother and baby survive

Systematic review

Systematic review of phaeochromocytoma in pregnancy

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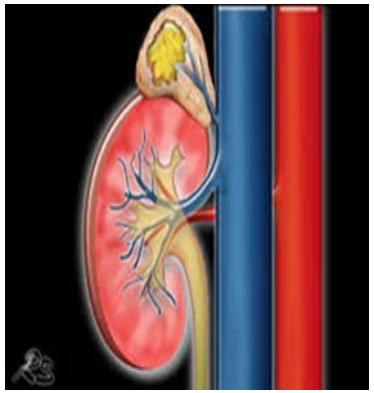

Conclusion:

- Phaeochromocytomas rare and dangerous condition in pregnancy
- Antenatal dxn reduces combined mortality from 50% to 15%
- Clues: classic triad, postural hypotension and absence of oedema and proteinurea
- Alpha blockade and fluids
- Early diagnosis – second trimester phaeo excision
- Late diagnosis – medical mx and concurrent or post partum resection
- Intrapartum diagnosis - medical and surgical emergency
- MDT of surgeons, endocrinologists, pharmacologists, anaesthetists and obstetricians