

# Interesting Case of Pediatric Hypertension

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# Case history

- 13 year old boy(Twin) from Erode, who was apparently normal 1 month before had fever for 3 days ,for which his parents consulted a general pediatrician.
- There incidentally boy was detected to have had hypertension.
- Blood pressure recorded was 180/130 mm hg in the rt upper limb.

# Causes of Pediatric Hypertension

## Causes of Childhood Hypertension According to Age Group

Age	Causes
One to six years	Renal parenchymal disease; renal vascular disease; endocrine causes; coarctation of the aorta; essential hypertension
Six to 12 years	Renal parenchymal disease; essential hypertension; renal vascular disease; endocrine causes; coarctation of the aorta; iatrogenic illness
12 to 18 years	Essential hypertension; iatrogenic illness; renal parenchymal disease; renal vascular disease; endocrine causes; coarctation of the aorta

# History relevant to HTN

- No history of abdominal pain, no history of oliguria/hematuria, no history of increased frequency of urine.
- No history of chest pain, paroxysms of palpitations, breathlessness, headache, sweating
- No history of arthritis, no history of rashes
- No history suggestive of weakness, muscle cramps, polyruia
- No history of drug intake
- No weight gain since 6 months, inspite of good appetite.

## History (Cont..)

- His twin sibling is normal.
- No similar complaints in the past
- No family history of hypertension.
- Paternal grandfather died of some chest tumor.

# On examination

- He got admitted in Apollo Hospital
- At admission,
- Weight: 41kg, Height: 165cm, normal BMI
- BP: upper limbs, supine – 140/90 mmHg, 108/min  
standing – 130/80 mmHg, 114/min,  
no radioradial or radiofemoral delay
- BP: lower limbs, 150/90mmHg
- Skin: no café au lait spots, no neurofibromas
- Systemic examination normal
- Medications:
  - Tab. Minipress XL 5mg OD
  - Tab. Amlodipine 5mg OD

# Pointers to hypertension

Physical findings	Potential relevance
<b>General</b>	
Pale mucous membranes, edema, growth retardation	Chronic renal disease
Elfin facies, growth retardation	Williams syndrome
Webbing of neck, wide spaced nipples, wide carrying angle, low hair line	Turner syndrome
Moon facies, buffalo hump, hirsutism, trunkal obesity, striae	Cushing syndrome
<b>Habitus</b>	
Thinness	Pheochromocytoma, renal disease, hyperthyroidism
Virilization	Congenital adrenal hyperplasia
Rickets	Chronic renal disease

# Pointers to hypertension

Skin	
Café au lait spots, neurofibromas	Neurofibromatosis, pheochromocytoma
Tubers, ash leaf spots	Tuberous sclerosis
Rashes	SLE, vasculitis
Palor, flushing, sweating	Pheochromocytoma
Needle tracks	Illicit drug use
Eyes	
Proptosis	Hyperthyroidism
Head and neck	
Goitre	Thyroid disease
Neurological signs	
Neurologic deficits	Chronic or severe acute hypertension with stroke



# Pointers to hypertension – systemic examination

Cardio vascular signs	
Absence or diminished femoral pulses, low leg pressure relative to arm	Aortic coarctation
Respiratory difficulty, hepatomegaly	Congestive heart failure
Bruit over great vessels	Arteritis or arteriopathy
Pulmonary signs	
Pulmonary edema	Congestive heart failure
Picture of Broncho pulmonary dysplasia	BPD associated hypertension

# Pointers to hypertension – systemic examination

Abdomen	
Epigastric bruit	Primary renovascular disease or in association with williams syndrome, neurofibromatosis, fibromuscular dysplasia, arteritis.
Abdominal masses	Wilms tumor, neuroblastoma, pheochromocytoma, polycystic kidney, hydro nephrosis
Genitalia	
Ambiguous, virilized	Congenital adrenal hyperplasia.

# History till now

- 13 yr
- Asymptomatic severe hypertension
- Normal BMI, no acanthosis nigricans
- No pointers to renal cause – history/clinical examination
- Normal difference between limbs (COA/aortoarteritis)
- No drug history

# Causes of Pediatric Hypertension

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# Investigations recommended

- Urine analysis
- Urine culture
- Serum creatinine
- Serum electrolytes
- Blood counts
- X ray chest and abdomen
- USG abdomen
- Intra venous urogram
- Voiding cystourethrogram

# Investigations recommended

- Radio nuclide imaging
- Selective renal angiography
- CT kidneys
- Doppler flow ultrasound
- MRI arteriogram
- CT angiography
- Renal vein assay
- Peripheral plasma renin activity/Plasma aldosterone
- Serum cortisol and 24 hour urine 17 hydroxy corticosteroid
- Urinary and plasma catecholamine levels
- Meta iodo benzyl guanidine ( MIBG) scan

# Our boy - In Erode

- Evaluated by Cardiologist in Erode
- Started on anti-hypertensive medications
- Chest X-ray showed a mediastinal lesion
- CT scan done in Erode: para-aortic mass – suspected as paraganglioma
- 24 hour urinary VMA was done – normal
  
- Referred to Chennai for Surgery
  
- Medications at admission:
  - Tab. Minipress XL 5mg OD
  - Tab. Amlodipine 5mg OD

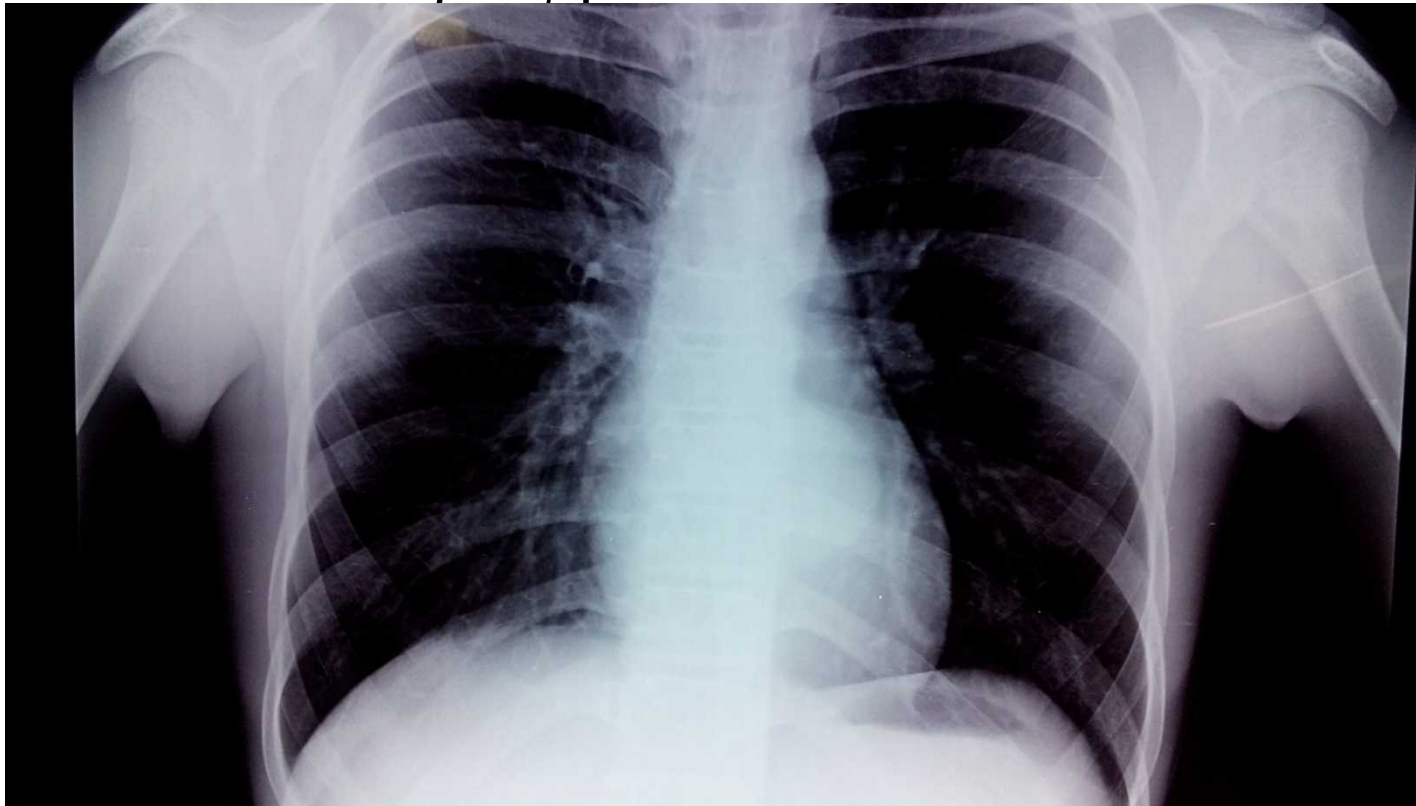
# Investigations done for our boy

- Urine routine examination normal
- Serum creatinine normal
- Serum electrolytes normal



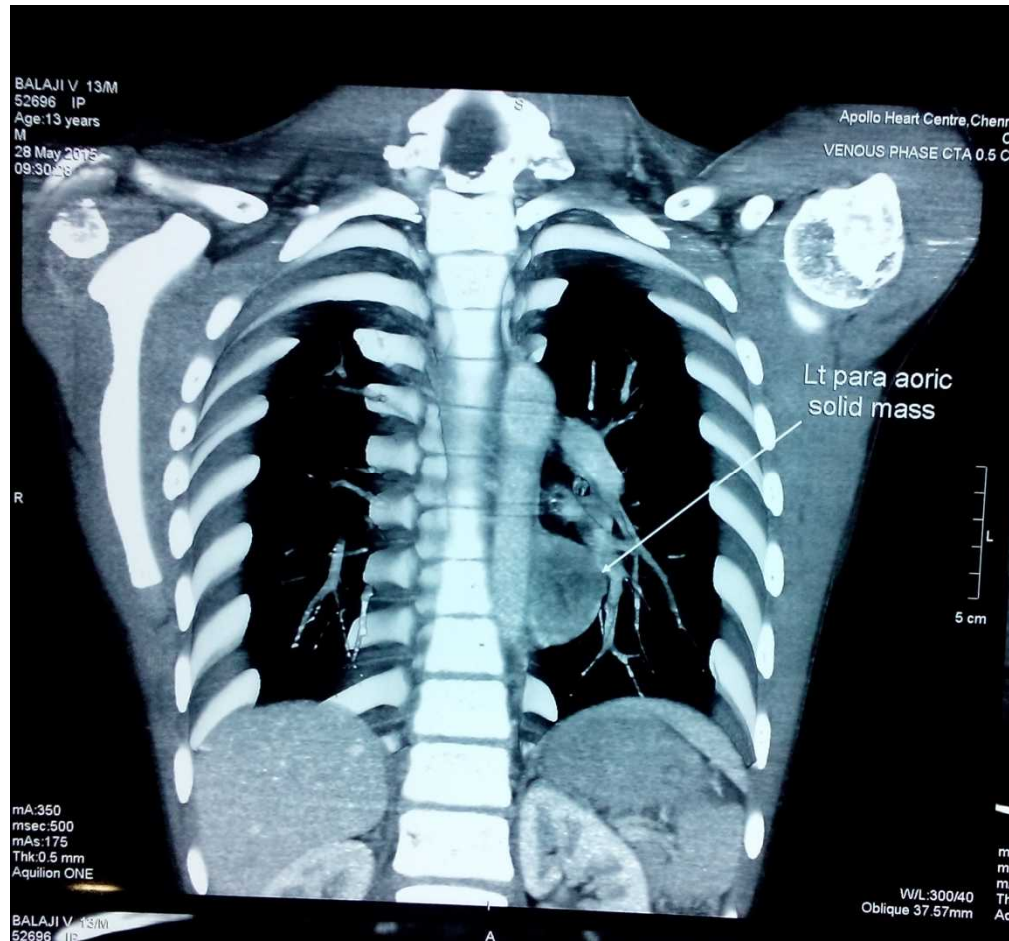
# Chest X-ray

- Well defined soft tissue lesion seen in the left retrocardiac region, paravertebral location



# CT Thorax

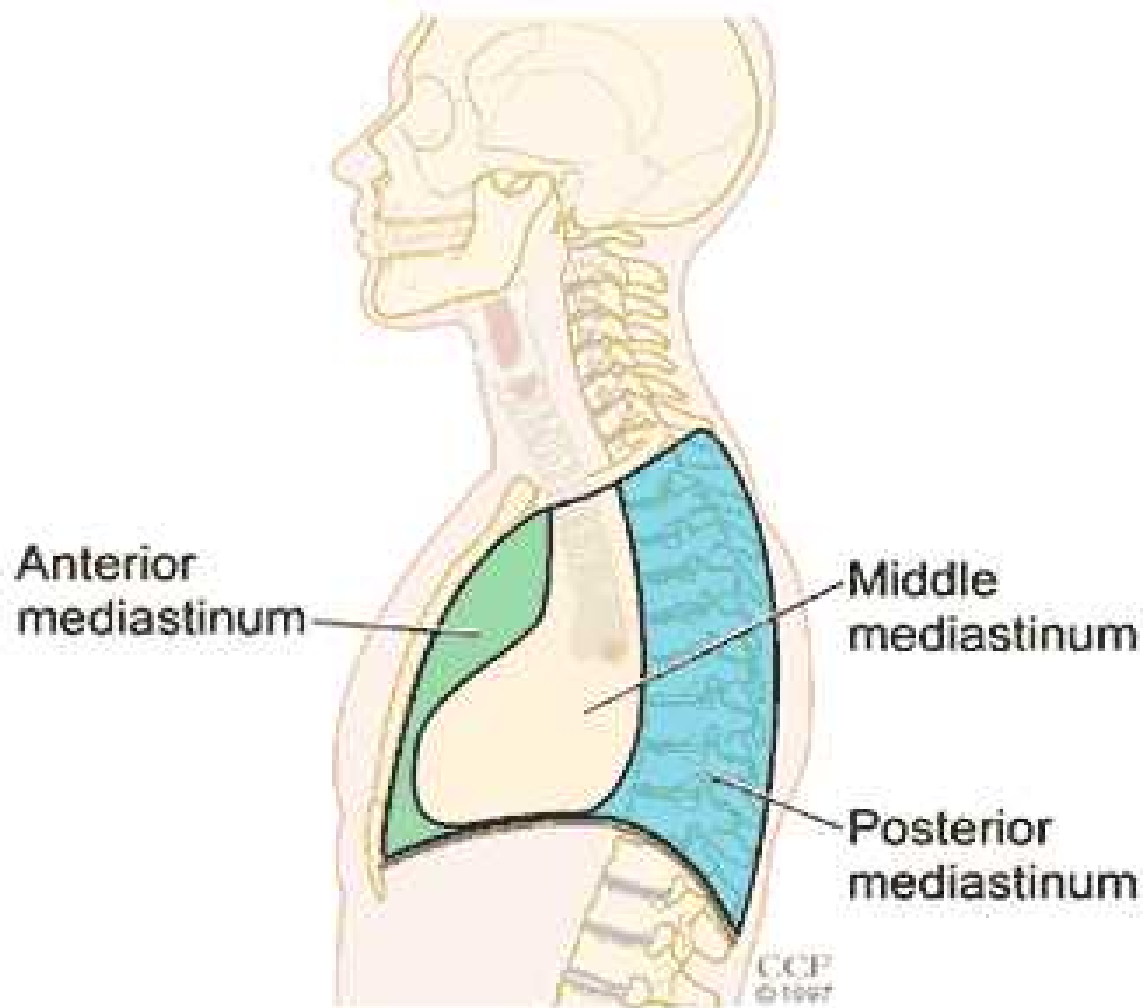
- 5 x 3 x 3 cm well circumscribed significantly enhancing solid mass in the para-aortic region
- D8-D10 levels







# Mediastinal masses



## POSTERIOR

- Neurogenic tumors
- Meningocele
- Gastroenteric cyst
- Mesenchymal tumors
- Pheochromocytoma
- Lymphoma

# Provisional Diagnosis

- Extra adrenal mediastinal pheochromocytoma (paraganglioma) with Hypertension

# Further Evaluation

- 24 hours urinary catecholamines
- I-131 MIBG Scan



# Catecholamines

- 24 hours urinary
  - Metanephrines: 180 mcg/24 hours ( 39 - 242)
  - Normetanephrines: 3784 mcg/24 hours ( 53 - 290)

# I-131 MIBG scan

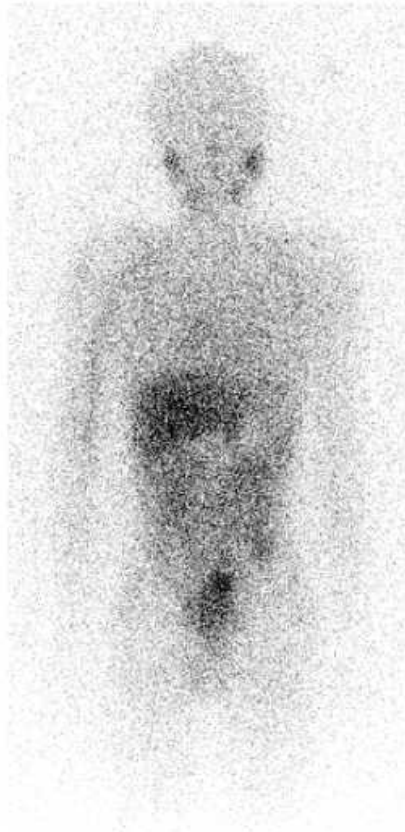
- Must if
  1. Suspected malignancy to know metastasis
  2. Bilateral pheochromocytoma
  3. Paraganglioma
  4. Youth with pheochromocytoma

# I-131 MIBG scan

Focal abnormal increased I-131 MIBG uptake noted in the left paravertebral region in the lower thorax

# 48 HRS IMAGING

I-131 MIBG WHOLEBODY 48 HRS



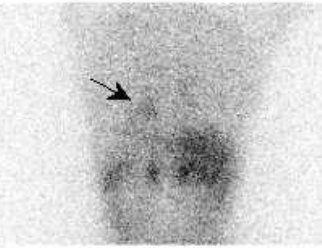
ANTERIOR  
72 HRS



POSTERIOR



ANTERIOR

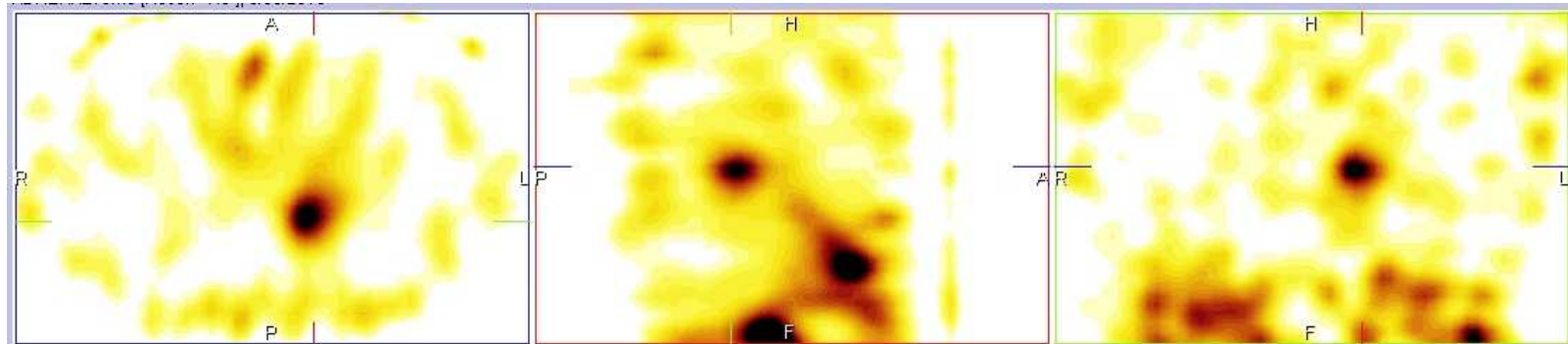


POSTERIOR

# I-131 MIBG/SPECT-CT

Focal uptake in the well defined soft tissue mass in left para-vertebral region at the D8-D10 level

# SPECT

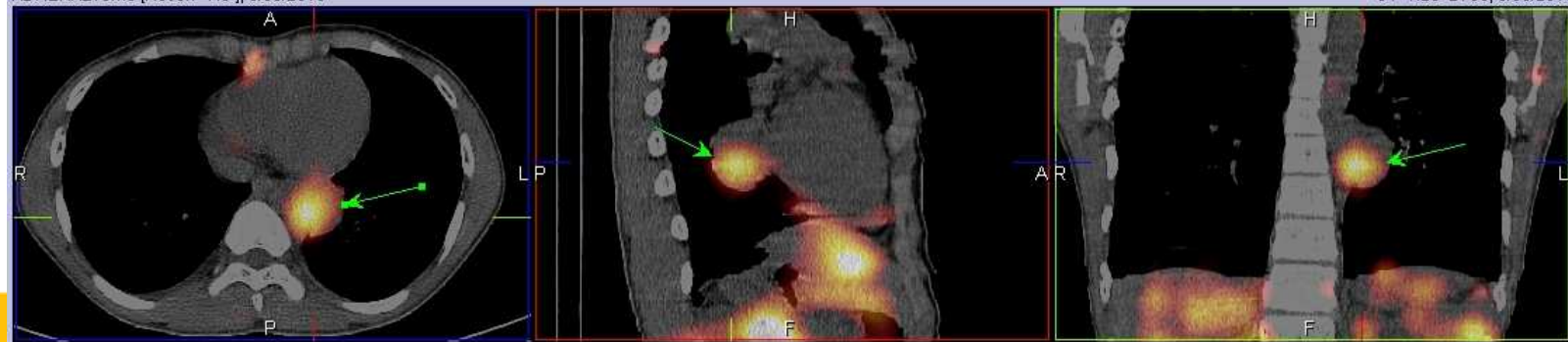


CT 1.25 B70s, 3/06/2015



ADRENALTomo [Recon - AC], 3/06/2015

CT 1.25 B70s, 3/06/2015



# Pre-operative preparation

- Volume expansion –
- Fluids
- Added Salt
- Control of BP/HR

First – alpha blockade

Do not use XL

After adequate alpha-blockade, beta-blockers to be added

At least of beta-blockers to be given for 3 days before surgery

CCB – Amlodipine can be added for better BP control

# Pre-operative preparation

- Encouraged adequate fluids up to 3L/day
- Added salt 10gm per day
- Minipress XL stopped and changed to plain Tab.prazosin 1.0mg TDS
- Tab.Atenolol 25mg once daily added
- Prior to Sx,
- Had BP controlled 110/74, 80min, 100/70, 84/min



# VATS excision of mediastinal tumor

VATS – video assisted Thoracoscopic surgery

By

- Dr.Rajiv Santosham
- Dr.Rajan Santosham

# Follow-up

- Postoperatively child bp is normalised in one day
- Biopsy report: specimen consistent with paraganglioma, posterior mediastinum.

# Post-op

- BP normal after 2 weeks of discharge
- Further plan
  - Repeat urine catecholamines after 1 month
  - Screening of twin sibling
  - Genetic analysis for familial paraganglioma- SDH gene, VHL gene planned
  - Life long follow up yearly

Abstract

Send to:

J Thorac Dis. 2014 Dec;6(12):1861-4. doi: 10.3978/j.issn.2072-1439.2014.12.30.

## Thoracoscopic resection of functional posterior mediastinal paraganglioma: a case report.

Ma L<sup>1</sup>, Mei J<sup>1</sup>, Liu L<sup>1</sup>.

Author information

### Abstract

A 48-year-old man with posterior mediastinal mass was diagnosed as functional mediastinal paraganglioma during surgical exploration via open thoracotomy in another hospital. The operation was terminated because of severe hypertension when touching the tumor. He was transferred to our center later. After systemic evaluation, the patient was medicated with oral alpha- and beta-blockades, as well as intravenous fluid resuscitation for two weeks. His blood pressure became stable and a second operation was planned. The tumor was removed completely via the thoracoscopic approach, and was finally confirmed as functional paraganglioma by immunohistochemistry. The patient recovered uneventfully after surgery, with no recurrence during one year follow-up visit.

**KEYWORDS:** Thoracoscopic resection; case report; functional mediastinal paraganglioma

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**Review** [Posterior mediastinal paraganglioma]. [Kyobu Geka. 2010]

Asymptomatic but functional paraganglioma of the posterior mediastinum [Ann Thorac Surg. 2014]

**Review** [Paraganglioma of the prostate: a case report and review]. [Zhonghua Nan Ke Xue. 2012]

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- First time – VATS to remove paraganglioma in a young age

# Take home message

- Importance of routine BP measurement even in younger age group
- In Pheochromocytoma/Paraganglioma
  1. Urine catecholamines -For confirming diagnosis
  2. MIBG Scan  
- For detecting other lesions
  3. Adequate alpha and beta blockade prior to surgery  
- For avoiding intra-operative crisis

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- Dr.Rajiv Santosham – Thoracic Surgeon
- Dr.Rochita – Radiologist
- Dr.Indrani – Nuclear Medicine Consultant
- Dr. Anruradha/Dr.Prabhu – Anaesthetist
- ICU Team
  
- But Who contributed the most for this Boy?

# Who contributed the most for this Boy?

- The pediatrician who checked BP in a child with fever





