# Child with bilateral pheochromocytoma and a surgically solitary kidney: Anesthetic challenges

*Manjunath Prabhu,*
*Tim Thomas Joseph,*
*Nanda Shetty, Souvik Chaudhuri*

*Department of Anesthesiology, Kasturba Medical College, Manipal, Karnataka, India*

**Address for correspondence:**
Dr. Tim Thomas Joseph,
Department of Anesthesiology,
Kasturba Medical College, Manipal,
Karnataka, India.
E-mail: timtjoseph@gmail.com

## ABSTRACT

Pheochromocytoma is a rare neuroendocrine tumor of childhood. We present a 14-year-old boy with bilateral pheochromocytoma, post nephrectomy in view of a non-functioning kidney presenting with severe hypertension and end organ damage. Diagnosis was confirmed with 24-hour urinary VMA, catechol amines, and CT scan. Preoperative blood pressure (BP) was controlled with prazosin, propranolol, nicardipine, and HCT-spironolactone. Anesthesia was given with general endotracheal anesthesia with epidural analgesia. Intraoperative BP rise was managed with infusion of NTG, MgSO4, esmolol, and dexmedetomidine which was especially challenging on account of bilateral tumor.

**Key words:** *Adrenalectomy, bilateral pheochromocytoma, solitary kidney*

## INTRODUCTION

Pheochromocytoma is a catecholamine-secreting neuroendocrine tumor that takes origin in the adrenal medulla or in chromaffin tissues along the paravertebral sympathetic chain.[1] It is most commonly present as paroxysmal spells of headaches, sweating, palpitations, and hypertension. Early diagnosis with aggressive, management is required for reducing mortality. Typically occurs in the age group of 30 to 50 years. The "rule of 10" has been described for pheochromocytomas. 10% of tumors are extra adrenal, bilateral, malignant, and occur in children. It is a rare tumor of childhood, more common amongst pre-adolescent boys with a greater tendency for familial occurrence.[2] The manifestations in children differ from those in adults. However, incidence of bilateral occurrence is higher in children (20%) than in adults.

## CASE REPORT

A 14-year-old boy, weighing 44 kg presented with complaints of excessive sweating since childhood, with recent onset fever, pedal edema, blurring of vision, easy fatigability, and Grade 2 dyspnea (NYHA). Four years ago, he underwent left nephrectomy in view of PUJ obstruction and a non-functioning kidney. His pulse rate was 102/min and blood pressure (BP) of 160/110 mm Hg. Adequate control of BP was achieved with tab. prazosin 7.5 mg tds, tab. nicardipine 40 mg bd, tab. propranolol 80 mg bd, and a combination of HCT-spironolactone 50 mg bd. A diagnosis of bilateral pheochromocytoma was made by raised levels of urinary catechol amines, ultrasound, and CT scan. CT scan showed bilateral adrenal masses with right adrenal mass measuring 5.8 × 4.1 × 4.3 cm and left mass was 6.4 × 2.9 × 3.8 cm [Figure 1]. 24-hour urine samples showed false-negative levels of HVA-14.3 mg/g (3-28 mg/g creatinine), VMA-8.6 mg/vol (2-8 mg/vol), but the ratio of VMA: Creatinine-38 mg/g of creatinine was elevated.[3] Creatinine level was 224 mg/vol (800-1 500 mg/vol). 24-hour urinary catechol amines was done which showed levels of adrenaline - 0.6 μg (<20 μg) and noradrenaline - 359.1 μg (<90 μg). Other laboratory parameters were within normal limits. Echocardiography showed an ejection fraction of 51%, concentric LVH, and fair LV systolic function. Renal artery Doppler was normal on right side. Ophthalmological evaluation showed Grade 4 hypertensive retinopathy, resolving papilledema and signs of optic atrophy. Child was posted for bilateral adrenalectomy following initial therapy with a BP of 140/80 mm Hg and no postural hypotension. Oral antihypertensives were continued as per schedule with preoperative anxiolysis. Maintenance

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| | DOI: 10.4103/1658-354X.114051 |

fluid was started overnight at 2 ml/kg/hr. Left radial artery was cannulated under local anesthesia. An epidural catheter was secured in right lateral position, in the T8-T9 space with careful positioning. 5 ml of 0.25% bupivacaine was administered. General anesthesia was induced using fentanyl 2 μg/kg and titrated doses of propofol. After adequate depth of anesthesia, gentle laryngoscopy was done and pharynx, epiglottis, and vocal cords were sprayed with 10% lignocaine. Right internal jugular cannulation was then performed (initial CVP was 15 mm of Hg). Intraoperative BP fluctuations during dissection and tumor manipulation of both sides were controlled with infusion of nitroglycerine (2-8 μg/kg/min) titrated to effect, $MgSO_4$ – 25 mg/kg, dexmedetomidine – 1 μg/kg, and boluses of esmolol 5-10 mg. Analgesia was provided with epidural infusion of 0.25% bupivacaine. IV hydrocortisone 100 mg was given. After excision of the second tumor, precipitous fall in BP was managed with an infusion of noradrenaline, dopamine, crystalloid, and colloid. Estimated blood loss was 300 ml. Urine output was maintained at 0.5-1 ml/kg/hr. Blood sugar was monitored. At the end of procedure, child was extubated. The gross specimen showed a single mass on the right and lobulated tumor on the left [Figure 2]. Postoperatively, child required noradrenaline 100 ng/kg/min for maintenance of BP temporarily. IV hydrocortisone 20 mg 8$^{th}$ hourly was started and tapered off in 18 days. Also, oral fludrocortisone 0.1 mg once daily was simultaneously put on. Child had uneventful postoperative period and was discharged on 12$^{th}$ postoperative day.

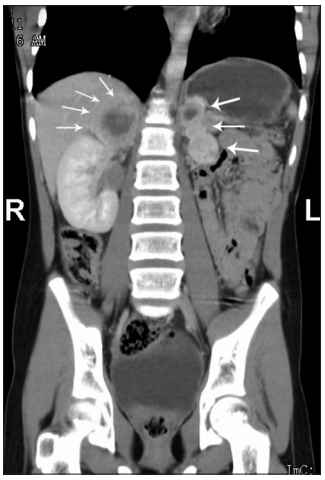


**Figure 1:** Bilateral adrenal masses with right adrenal mass measuring 5.8 × 4.1 × 4.3 cm and left lobulated mass 6.4 × 2.9 × 3.8 cm

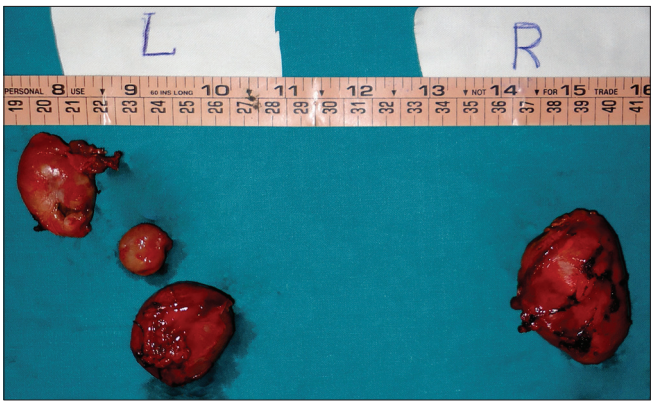


**Figure 2:** Gross specimen showed a single mass on the right and lobulated tumor on the left

## DISCUSSION

Adrenal pheochromocytoma secretes adrenaline and noradrenaline, whereas extra-adrenal tumors secrete only noradrenaline. Usually, the secretion of noradrenaline is greater than adrenaline.[4] It is seen more commonly among pre-adolescent boys and teenage girls, with a male preponderance (2:1).[5,6] The clinical picture in children differ from adults (9%). The incidence of bilateral disease is higher in children (20%). The classical symptoms of pheochromocytoma are headaches, hypertension, palpitations, and episodic sweating. Hypertension due to pheochromocytoma in children tends to be more severe and refractory than in adults.[2] The patients presenting with pheochromocytoma crisis may have extremely varied manifestations, ranging from severe hypertension to circulatory failure and shock.[7] In our case, child had developed features of end organ damage like hypertensive retinopathy, optic atrophy, and cardiac dysfunction. Diagnosis is usually confirmed by raised urinary catecholamine levels and their metabolites. VMA gives 15% false results and measurement of 24-hour free urinary catecholamine is the best confirmatory test which was necessary in our case. Localization of tumor is accurately done by CT scan, MRI, and MIBG.[8] Preparation for surgery includes pharmacological control of adverse effects of circulatory catecholamines and restoration of blood volume by appropriate α blockade and achieve adequate BP control over a period of 10 to 14 days. β blockers are necessary in cases of significant dysrhythmias or tachycardia.[9] Catecholamines cause constriction of arteriolar and venous segments and thereby decreasing the circulating blood volume.[10] Goals of anesthetic management aim at providing optimal surgical conditions and suppress the responses to endotracheal intubation, surgical stimulation, tumor handling, and revascularization. Epidural anesthesia with general anesthesia helped increase vascular capacity and analgesia. During tumor manipulation and resection, we used NTG and MgSO4 BP fluctuations. Esmolol and dexmedetomidine were found effective for heart rate control. A solitary kidney mandates extra caution,

which is also prone to injury intraoperatively. In such cases, avoid dehydration, maintain renal perfusion, and avoid nephrotoxic agents. Glucocorticoid and mineralocorticoid cover is mandatory for patients undergoing bilateral resection. Preparedness to tackle inevitable drop in pressures after complete excision of tumor with vasopressors is necessary. In bilateral tumors, the hemodynamic fluctuation are more pronounced and prolonged as there are two endocrine tumors to excise. Contracted vascular compartment with volume depletion warrants prompt resuscitation with crystalloids and colloids. To conclude, prompt and early diagnosis, meticulous planning, and a multidisciplinary approach are quintessential in reducing the mortality and morbidity associated with pheochromocytoma in children.

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**How to cite this article:** Prabhu M, Joseph TT, Shetty N, Chaudhuri S. Child with bilateral pheochromocytoma and a surgically solitary kidney: Anesthetic challenges. Saudi J Anaesth 2013;7:197-9.
**Source of Support:** Nil, **Conflict of Interest:** None declared.