Recent Trends in the Perioperative Management of Pheochromocytoma

An Essay

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List of Abbreviations

¹⁸FDA 18-fluorodopamine

¹⁸F-DOPA 18-fluoro-dihydroxyphenylalanine.

ACTH Adrenocorticotropic hormone.

AD Aldehyde Dehydrogenase.

AVP Arginine Vasopressin.

AVS Adrenal venous sampling.

BMI Body mass index.

CGA Chromogranin A.

CI Cardiac index.

COMT Catechol O-Methyltransferase.

CT Computed tomography.

DBH Dopamine Beta Hydroxylase.

List of Abbreviations

DDC Dopa decarboxylase.

DHEA Dehydroepiandrosterone.

DHPG 3,4-Dihydroxyphenylglycerol.

Dopa 3,4-dihydroxyphenylalanin.

DOPAC 3,4-dihydroxyphenylacetic acid.

ECG Electrocardiography.

HCT Hematocrite.

HPLC High-performance liquid chromatography.

HVA Homovanillic Acid.

IAP Intra-abdominal pressure.

ICU Intensive care unit.

ISP International Symposium on Pheochromocytoma.

IVC Inferior vena cava.

LA Laparoscopic adrenalectomy.

LC-ED Liquid chromatography with electrochemical detection.

LCMS/ MS Liquid chromatography with tandem mass spectrometry.

MAO-A Monoamine oxidase A.

MB Methylene blue.

MEN Multiple endocrine neoplasia.

MHPG Methoxy Hydroxyl Phenyl Glycerol.

MIBG ¹²³I-meta-iodo-benzyl-guanidine.

MRI Magnetic resonance imaging.

MSCT Multislice spiral computed tomography.

NF-1 Neurofibromatosis type I.

E List of Abbreviations

PACU Post-anesthesia care unit.

PASS Pheochromocytoma of the Adrenal Gland Scaled Score.

PET Positron emission tomography.

PET-CT Positron emission tomography.

PG Pico gram.

PNMT Phenylethanolamine N-methyltransferase.

PPG Pheochromocytoma and extra-adrenal paraganglioma.

PRA Posterior retroperitoneoscopic adrenalectomy.

PTHRP Parathyroid hormone-related peptide.

SDH Succinate dehydrogenase.

SIA A syndrome of inappropriate anti-diuretic hormone.

SUV Standardized uptake value.

E List of Abbreviations

Th Tyrosin hydroxylase.

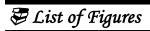
VHL Von Hippel-Lindau.

VIP Vasoactive intestinal peptide.

VMA Vanillylmandelic acid.

VMAT1 Vesicular monoamine transporter 1.

VMAT2 Vesicular monoamine transporter 2.



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Introduction

Pheochromocytoma is a rare tumor arising from chromaffin cells in adrenal medulla or other paraganglia in the body, which may be associated with many genetic syndromes and mutation (*Mackenzie et al.*, 2007).

Pheochromocytoma has got multidimensional challenging aspects in spite of our improved understanding of its physiological and clinical behavior during surgical resection. This neuroendocrine tumor is associated with a most unpredictable and fluctuating clinical course during anesthesia and surgical intervention. The difficulties and challenges increase manifold in patients with undiagnosed accidental diagnosis or pheochromocytoma who present to the hospital for the treatment of some other disease or emergency (Mackenzie et al., 2007).

The perioperative management of pheochromocytomas requires meticulous anesthetic care. Although there has been considerable progress in its management, recent progress has defined 4 agents sulphate, clevidipine, (magnesium vasopressin methylene blue) that may be particularly advantageous to the anesthetic team (Lord and Augoustides., 2012).

Anatomy of the Adrenal Gland

The adrenal (supra renal) glands are two small yellowish organs, flat antero-posteriorly and lying immediately antro superior and medial to each upper renal pole (figure 1), they lay within perinephric fat and enclosed in the renal fascia but separated from the kidneys by fibrous tissue (*Standering*, 2008).

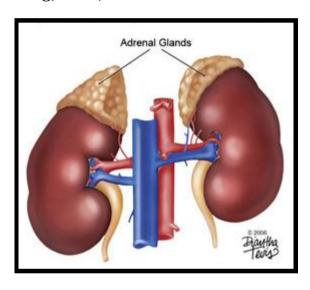


Figure1: Kidneys and adrenals (Standering, 2008).

Anatomy

• <u>Gross anatomy</u>: The adrenal gland is approximately one third the size of the kidney at birth. However, in the adult, the adrenal gland is only one thirtieth the size of the kidney. This change in proportional size is a reflection of renal growth as well as the involution of the fetal adrenal cortex after birth.

At the onset of puberty, the gland is at its adult size and only increases slightly in weight over the course of adult life, with the exception of stress and pregnancy or the development of pathology. A normal adrenal gland in an adult weighs approximately 4 to 6 grams. The left adrenal is larger and flatter than the right adrenal gland. The weight of each adrenal may increase by nearly 50 percent during times of stress and pregnancy. Pathologic glands may reach 700 grams (*Kebebew and Duh.*, 2001).

The normal adrenal cortex and medulla have different characteristics:

- The normal adrenal cortex is dark yellow and has a firm consistency and finely granular surface, allowing it to be differentiated from surrounding adipose tissue. The cortex composes 80 to 90 percent of the volume of a normal gland.
- The central medulla is red-brown and is enclosed completely by the adrenal cortex, except at the hilum. The medulla constitutes 10 to 20 percent of the volume of a normal gland (*Avisse et al.*, 2000).

The medulla produces catecholamines (adrenaline, noradrenaline) as well as dopamine. The cortex is subdivided into three zones (from outer to inner):

- 1. Zona glomerulosa.
- 2. Zona fasciculate.

3. Zona reticularis (Elsayes et al., 2004).

The zona glomerulosa is responsible for the production of mineralocorticoids, mainly aldosterone, which regulates blood pressure and electrolyte balance (*Elsayes et al., 2004*).

The zona fasciculata, is responsible for the production of glucocorticoids, predominantly cortisol, which increases blood sugar levels via gluconeogenesis, suppresses the immune system, and aids in metabolism. This zone secretes cortisol both at a basal level and as a response to the release of adrenocorticotropic hormone (ACTH) from the pituitary gland (*Elsayes et al., 2004*).

The zona reticularis produces gonadocorticoids and is responsible for administering these hormones to the reproductive regions of the body. Most of the hormones released by this layer are androgens. The main androgen produced by this layer is dehydroepiandrosterone (DHEA), which is the most abundant hormone in the body and serves as the starting material for many other important hormones produced by the suprarenal gland, such as estrogen, progesterone, testosterone, and cortisol (*Elsayes et al.*, 2004).

• Extra-adrenal gland anatomy

True accessory adrenal glands, which contain both cortical and medullary tissue, are rare. Most extra-adrenal glands contain either cortical or medullary tissue.