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PARTIAL ADRENALECTOMY FOR FAMILIAL AND HEREDITARY PHEOCHROMOCYTOMA

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Traditionally total adrenalectomy has been advocated for the treatment of bilateral adrenal disorders especially in cases of hereditary syndromes like multiple endocrine neoplasia type 2, Von Hippel–Lindau disease and neurofibromatosis type I. However, currently it is well recognized that total adrenalectomy is associated with the morbidity of medical adrenal replacement therapy. Lifelong adrenal replacement therapy after bilateral adrenalectomy may predispose patients to osteoporosis, Addisonian crisis and decreased quality of life. In that context, partial adrenalectomy has been suggested for patients with functioning and non-functioning benign adrenal tumours especially in the case of hereditary adrenal-producing syndromes, bilateral or multifocal lesions or solitary adrenal glands. Advantages of partial adrenalectomy include preservation of adrenocortical function and catecholamine excretion while resultant avoidance of post-operative chronic steroid replacement.

Cortical sparing adrenalectomy has been described in both hereditary and sporadic pheochromocytoma. The use of cortical sparing adrenalectomy is highly debated in the case of unilateral pheochromocytoma due to the difficulty in excluding malignancy. The majority of literature data focus on hereditary pheochromocytoma patients with RET or VHL mutations. The low risk of malignancy and high risk of bilateral tumours are obvious in the above genetic syndromes especially in MEN2. Recurrence rate is estimated at about 10% for pheochromocytoma. Overall steroid dependence rate is estimated at 90%. Long term follow up of the patients has not been standardized. The surgical technique has not been standardized and open questions remain regarding the tumour margin, the adrenal vein preservation, the means of haemostasis. The lecture will focus on the indications of partial adrenalectomy in the case of familial and hereditary pheochromocytoma, surgical technique, on complications as well as on outcome.

BIOGRAPHY

Yiannakopoulou E is an endocrine surgeon, Breast surgeon and pharmacologist. Current position: Faculty in the Department of Biomedical Sciences, Faculty of Health and Caring Professions, University of West Attica, Athens, Greece; Director of her own private medical practice at University of Strasbourg, France.

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