Treatment

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Medical Metyrapone or ketoconazole therapy reduces steroid synthesis and secretion by CYP11B1 inhibition and can be used to prepare patients with severe hypercortisolism preoperatively or as primary therapy if surgery is not possible. In patients who are critically ill with Cushing’s syndrome, intravenous etomidate infusion (even at non-hypnotic doses) can reduce serum cortisol levels to normal within 24 hours, providing a suitable window for surgery . This requires monitoring in an intensive care unit setting. Surgical ACTH-producing pituitary tumours are treated by trans - sphenoidal resection and/or radiotherapy . Resection of ectopic ACTH-secreting tumours will also correct hypercortisolism. Patients who have undergone failed pituitary surgery or those with an unresectable or unlocalised ectopic ACTH-secreting tumour may require bilateral adrenalectomy to control hormone excess. This will render them steroid dependent. Unilateral adenoma should be treated by minimally invasive adrenalectomy provided adrenocortical cancer is not suspected. In cases of bilateral ACTH-independent disease (Figure 57.6), the extent of adrenalectomy is contentious; bilateral adrenalectomy should be employed in severe Cushing’s and equally enlarged adrenals. In asymmetric disease, excision -

Figure 57.6 Bilateral asymmetrical hyperplasia of the adrenal glands (arrows) in a patient with Cushing’s syndrome.

In cases of subclinical Cushing’s syndrome and a unilateral adenoma, unilateral adrenalectomy is indicated if the tumour is >4 /uni00A0 cm or <4 /uni00A0 cm with features of the metabolic syndrome. Cushing’s syndrome predis poses patients to increased risk of venous thromboembolism, cardiac events, infection and poor wound healing. Patients should therefore receive chemical and mechanical thrombo prophylaxis as well as perioperative broad-spectrum antibiotics. Accompanying diabetes should also be adequately controlled. As unilateral or bilateral adrenalectomy in the setting of Cushing’s syndrome will result in steroid deficiency in the postoperative period, patients should r eceive intraoperative corticosteroids (50–100 /uni00A0 mg intravenous hydrocortisone) and close liaison with the endocrinology team is strongly advised to guide postoperative management. After unilateral adrenalectomy the contralateral gland will be suppressed and so all patients should be commenced on 15–25 /uni00A0 mg daily of hydrocortisone. In total, 15 /uni00A0 mg/h is requir ed parenterally for the first 12 hours followed by a daily dose of 100 /uni00A0 mg for 3 /uni00A0 days, which is gradually reduced thereafter. After unilateral adrenalectomy , the contralateral suppressed gland may need up to a year to recover adequate function. A synacthen test is used to confirm adequate adrenal function prior to stopping hydrocortisone supplementation. In 10% of patients with Cushing’s disease who undergo a bilateral adrenalectomy after failed pituitary surgery , the pituitary adenoma causes Nelson’s syndrome owing to continued ACTH secretion at high levels, resulting in hyperpigmentation due to uncontrolled secretion of pro-opiomelanocortin (POMC). POMC is cleaved to produce ACTH and melanocyte-stimulating hormone, excess of the latter resulting in hyperpigmentation.

Preoperative management. Postoperative management.

Revision #1
Created 2025-12-31 15:21:32 UTC by Omar Ayman
Updated 2025-12-31 15:21:32 UTC by Omar Ayman