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Update on Adrenal Disorders

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Abstract

The spectrum of adrenal disorders in childhood is wide and thorough clinical history and examination and appropriate investigations can facilitate accurate diagnosis and management.

Congenital adrenal hyperplasia (CAH) is a common condition with nuances in the presentation which can point towards different diagnoses. Simple virilising CAH in a boy can present with gonadotropin independent precocious puberty caused by excessive adrenal androgen production. The puberty can progress on treatment as the high androgen drive primes the hypothalamic pituitary gonadal axis and can trigger gonadotropin dependent precocious puberty which will require GnRH analogue treatment. Long term sequelae of CAH treatment can be affected by poor compliance with therapy. Care should be taken to diagnose testicular adrenal rest tumours as they may compromise fertility.

Glucocorticoid resistance is a rare condition of partial end-organ insensitivity to glucocorticoids, with compensatory elevation of ACTH and Cortisol. The excess ACTH can also cause mineralocorticoid excess and hyperandrogenism. This

can cause a variety of clinical presentations including signs of mineralocorticoid excess, and androgen excess, and early and excessive adrenarche can be associated with precocious puberty. Males can present with abnormal spermatogenesis. It is important to consider this important condition in the differential diagnosis of adrenarche and endocrine hypertension especially if the cortisol and ACTH levels are raised.

Pediatric Cushing's syndrome is uncommon and appropriate investigation is needed to distinguish between pituitary from adrenal forms. Medical management can alleviate some of the symptoms but surgical removal of the pituitary adenoma is indicated. If the removal is incomplete some patients will need radiotherapy and ongoing medical treatment. Rarely, adrenalectomy may be indicated in refractory cases of Cushing's syndrome.

In this talk, various clinical scenarios will be presented to highlight the spectrum of adrenal disorders.



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