

Handling of internal contamination

It is the responsibility of AGRC members to advise on the removal of radionuclides from the respiratory and gastrointestinal tracts and the management of radionuclides absorbed into the body. Physical removal by nose blowing, irrigation, mouth washes, removal by swabbing, etc. are the usual methods. Nasal and mouth swabs, urine and faeces are repeatedly monitored where necessary and at times the chest and whole body are scanned. Certain manipulations may become necessary to counter internal contamination, such as the giving of products to lower absorption from the gastrointestinal tract, limit biological activity and increase excretion rates.

Concluding phase

After decontamination and surgical management the patient is removed to a non-contaminated clean zone. The nurse in

the neighbouring clean zone will roll a 1,5 m wide plastic floor covering into the controlled zone next to the table on which the patient lies. A clean trolley will be introduced for transferring the patient from the operating theatre. All personnel in the controlled zone remove their gowns, gloves, etc. and are monitored for residual radioactivity before leaving the area. All covering, clothing, instruments, towels, etc. used in the theatre are labelled for subsequent examination by the physicists and handled according to their recommendations. Before leaving the TRCF all staff are monitored for local contamination.

Postoperative period

This period can be complicated by the manifestation of the acute radiation syndrome; careful monitoring and teamwork are therefore required. The AGRC is especially involved in supporting the surgeon. The many facets of the postoperative phase are beyond the scope of this article.

Kallmann's syndrome with unilateral renal agenesis

A case report

S. HONIBALL, M. SANDLER

Summary

A case of Kallmann's syndrome (hypogonadotropic eunochoidism plus anosmia) in which further investigation revealed the association of unilateral renal agenesis is described. The importance of excretory urography in the investigation of patients with Kallmann's syndrome is stressed.

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The association of underdeveloped genitalia and the absence of olfactory lobes was first described in 1856 in an autopsy report.¹ In 1944 Kallmann described the original syndrome in which reduced luteinizing hormone (LH) and/or reduced follicle-stimulating hormone (FSH) was found in association with hyposmia or anosmia, syndactyly, short fourth meta-

carpals, colour blindness, nerve deafness and mental retardation.² Since the original description other congenital anomalies have been reported in Kallmann's syndrome including harelip,³ cleft palate,⁴ and cryptorchidism.⁵

A patient in whom investigations revealed the rarely observed association of unilateral renal agenesis with Kallmann's syndrome is described.

Case report

A 28-year-old man was referred for investigation of delayed puberty and cryptorchidism. The history revealed the absence of palpable testes, which had occasionally been felt to descend into the inguinal canal. At puberty there had been scanty development of pubic and axillary hair. On specific questioning the patient acknowledged a poor sense of smell, but denied the presence of similar findings or history of infertility in other family members; however, the patient's 12-year-old nephew had impalpable testes. Physical examination showed a tall man (height 1,72 m, span 1,79 m) with a female habitus and sparse facial and axillary hair. The cardiovascular, respiratory and gastro-intestinal systems were normal. CNS examination showed a man with normal intellect, normal hearing but an impaired sense of smell on quantitative testing. Examination of the urogenital system revealed minimal pubic hair, an infantile penis and bilateral impalpable testes.

The presence of hypogonadism, cryptorchidism plus anosmia were highly suggestive of Kallmann's syndrome. This diagnosis was supported by the findings of low serum LH, FSH and testosterone levels on three separate occasions, in the presence of normal serum biochemistry and hormone profiles (Table I). Combined pituitary testing with Actrapid insulin (Novo) 0,15 µg/kg,

Endocrine Unit, Department of Internal Medicine, University of Stellenbosch and Tygerberg Hospital, Parowvallei, CP

S. HONIBALL, M.B. CH.B.

M. SANDLER, M.B. CH.B., M.R.C.P.

Reprint requests to: Dr M. Sandler, Dept of Internal Medicine, University of Stellenbosch, PO Box 63, Tygerberg, 7505 RSA.

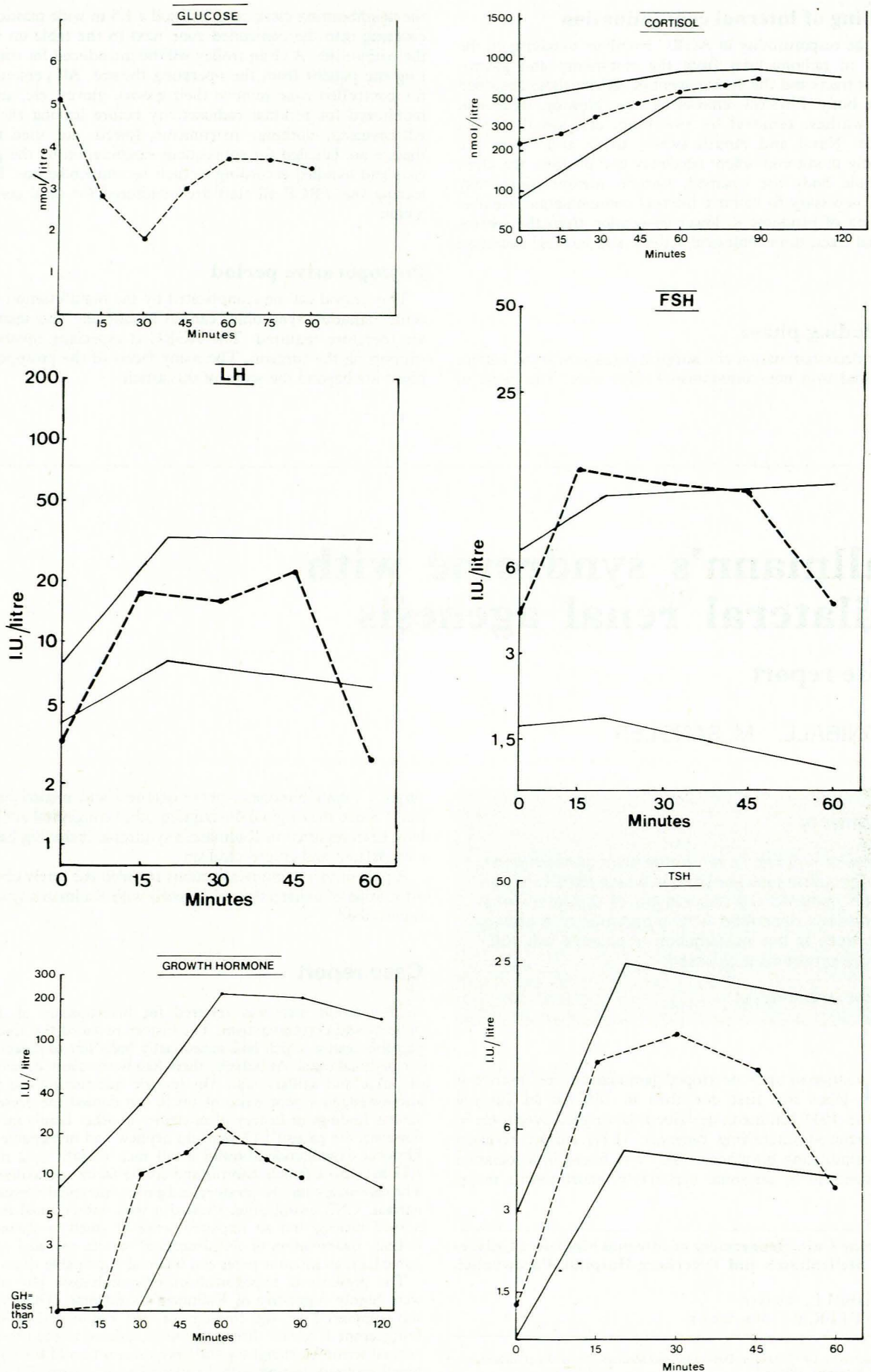


Fig. 1. Insulin hypoglycaemia with GnRh and TRH stimulation (combined pituitary test) showing essentially intact pituitary function (solid lines represent the upper and lower limits of normal reference values for our laboratory; broken line represents the patient's measured data).

TABLE I. BASELINE SERUM BIOCHEMISTRY AND HORMONE PROFILES

	Patient	Normal values*
Luteinizing hormone (U/l)	2,9 - 3,3	5 - 50
Follicle-stimulating hormone (U/l)	2,6 - 4,3	5 - 20
Thyroid-stimulating hormone (mU/l)	1,2	5,0
Prolactin (μ g/l)	5,0	0 - 20
Cortisol (nmol/l)	664,0	193 - 690
Free thyroxine (pmol/l)	13,9	8,8 - 23,0
Free tri-iodothyronine (pmol/l)	2,3	3,0 - 8,6
Urea (mmol/l)	3,8	3,3 - 6,5
Creatinine (μ mol/l)	90,0	60 - 120

*Normal reference values for our laboratory. Hormones were determined by standard radio-immunoassay.

gonadorelin (H.R.F.; Ayerst) 100 μ g, and Relefact thyrotrophin-releasing hormone (TRH) (Hoechst) 200 μ g showed initial low basal gonadotrophins which responded normally to stimulation in the presence of further intact pituitary function (Fig. 1). Investigations for the presence of previously documented associations with Kallmann's syndrome included normal visual acuity and a normal skeletal survey and audiogram. An excretory urogram revealed renal agenesis on the right with a normally functioning left kidney.

The patient was treated with human chorionic gonadotrophin (HCG) (Profasi; Serono) 2000 U intramuscularly twice weekly for 6 months. The response to therapy was only partial in that pubic hair developed, both testes became palpable while the penis and scrotum remained underdeveloped and the serum testosterone value was 1,6 nmol/l (normal — 8,65 - 29,5 nmol/l). Treatment with long-acting testosterone (Sustanon-250; Organon) 1 ml intramuscularly once a month was commenced. After 6 months' therapy there was a marked clinical improvement in that both pubic and axillary hair had increased and the penis had attained a normal length.

Discussion

Delayed puberty is a common disorder, which is often belatedly diagnosed and treated. A poor sense of smell is rarely volunteered by these patients and it is therefore important to ascertain this fact specifically during history-taking and then

to confirm its presence or absence by quantitative testing. The presence of an abnormal sense of smell can help to differentiate Kallmann's syndrome from constitutionally delayed puberty — this differentiation being one of the most difficult to make in clinical endocrinology. The clinical diagnosis of Kallmann's syndrome in this case was supported biochemically by the presence of hypogonadotropic hypogonadism. The gonadotrophins responded normally to gonadotrophin-releasing hormone (GnRH) stimulation indicating that the lesion was hypothalamic in origin, a finding confirmed by previous reports.⁴ The response to treatment of patients with Kallmann's syndrome is not usually affected by the presence of the associated phenomena, except in the presence of bilateral cryptorchidism — where the response to HCG therapy has been noted to be incomplete,⁶ a finding confirmed in our patient.

The association of Kallmann's syndrome with unilateral renal agenesis has been previously described in only 5 patients.^{5,7,8} Although the occurrence of this association seems rare, it is nevertheless important to consider this possibility when investigating a patient suspected of having Kallmann's syndrome, and we therefore suggest that excretory urography be included in the routine investigation of such a patient.

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