likelihood of elevated PCWP would be higher than in the "general population" usually referred for GCBP study. The remaining ten patients also appear to have higher likelihood of elevated PCWP, since six of them had cardiomyopathy; one had a myxoma, one VSD, and two ASD. Prevalence of PCWP greater than 12 mm Hg in the whole group was 37%.

The sensitivity of 81% and specificity of 89%, when applied to a population with 37% prevalence of the condition (elevated PCWP) yields a predictive value of 81% for a positive result and 89% for a negative result. However, if a prevalence of 10% is assumed, the predictive value of a positive result will be 45%, and for the prevalence of 5%, the predictive value will be 28%.

The cardiac blood-pool study by virtue of its noninvasive nature and the information it supplies is considered a clinically valuable test. However, its value depends on the spectrum of the conditions it is used for, and is limited to a "selected group" of patients in whom a specific question can be, or is expected to be, answered by this test with a certain degree of confidence.

Regarding the clinical usefulness of the Urbina et al. finding (A/B ratio and its correlation with PCWP), this specific question may be asked only in a small percentage of patients referred for GCBP study, in whom, and in these, if the result is positive, the degree of confidence would be too low to give the test any clinical value.

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## Reply

Thank you for your comments regarding the referenced article on capillary wedge pressure as related to blood-pool studies. Your statements about the importance of the population under investigation are quite relevant to the value of any procedure. However, the primary goal of Dr. Urbina's report was to establish the relationship between the pulmonary capillary wedge pressure, as determined in the catheterization laboratory, and apex/base ratio as determined from the blood-pool scan. It is impressive that even though many of these measurements were not made simultaneously, the apex/base ratio was able to differentiate between normal and abnormal pressures *in the group under investigation* with a sensitivity and specificity as reiterated in your letter.

Certainly, the data reported by Urbina should be checked in other laboratories. When other series of patients are investigated, it is possible that groups with a lower prevalence of disease will be selected for study. Sensitivity and specificity will probably be different in those circumstances. The importance of Dr. Urbina's study, however, lies in its utilization of data that are already present on the blood-pool scan to obtain further insight into the physiologic state of the patient under investigation.

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## Co-existing Klinefelter's Syndrome, Sublingual Thyroid, and Hypothyroidism

A case of co-existing Klinefelter's syndrome (KS), sublingual thyroid, and primary hypothyroidism is presented.

A 24-year-old male electrician was referred for evaluation of infertility, tiredness, and obesity. He had been married 2 yr but was unable to beget a child, in spite of normal libido and sexual function. Past history revealed that he was given thyroid hormone pills for "sluggishness" intermittently by his physician. He had not taken any medication for several years before this evaluation.

Physical examination revealed a large, obese Caucasoid male, 180 cm tall and weighing 127 kg. The skin was normal except for sparse facial and pubic hair. Examination of the neck revealed no palpable thyroid or nodules. The testes were very small and the penis was normal to small in size. The neurologic examination, including reflex relaxation, was within normal limits.

The iodine-123 thyroid scan revealed an unsuspected sublingual thyroid but no radioiodine uptake in the usual thyroid bed (Fig. 1). The 24-hr radioiodine uptake was 13% (nl 10-35%), measured with the uptake probe centered over the sublingual area. Retrospective examination of this area revealed a barely palpable mass above the hyoid. Thyroid function tests revealed serum T4 of 5.2 μg/dl (nl, 5.4-12.3); T3 uptake ratio of 0.96 (normal pooled serum = 1.00); free T4 index of 5.0 (nl, 5.4-12.3); and a TSH level of 19.3 microU/ml, (nl 1.0 to 6.2), which confirmed the primary hypothyroidism.

Other pertinent laboratory findings were: elevated serum follicle-stimulating hormone (FSH), 70 uIU/ml (nl, 4-25), and luteinizing hormone (LH), 40 uIU/ml, (male nl 7-24); decreased plasma testosterone, 78 ng/dl (nl 300-800); absence of spermatozoa on semen sample; and positive X chromatin on buccal smear. The diagnoses of Klinefelter's syndrome, sublingual thyroid, and primary hypothyroidism were made. Thyroid hormone and testosterone replacement was suggested, and the patient was returned to his local doctor.

In patients with KS, co-existing hypothyroidism has been reported sporadically (*1-5*). Many investigators have tried to find a possible underlying disorder in the hypothalamo-pituitary-thyroid axis in patients with KS.

Davis et al. (*6*) studied five patients with KS. Three of them had decreased radioactive iodine uptake (10%, 9%, 8%). The TSH-stimulated radioiodine uptakes were 40%, 47% and 17% respectively. This suggested a possibility of pituitary insufficiency, but

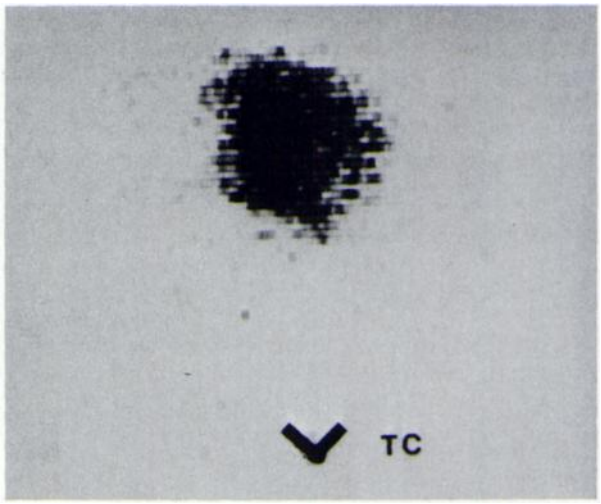


FIG 1. I-123 neck Scan. Note sublingual thyroid and lack of activity in usual thyroid bed. TC = thyroid cartilage.

not primary thyroid abnormalities. With the use of the TRH stimulation test, some authors have found a blunted TSH response in patients with KS (*7–8*). Possible explanation for this phenomenon was that the chronic overproduction of gonadotropins could result in a decreased availability of the alpha-sub unit required for biochemical synthesis of TSH (*9*). On the other hand, Burman et al. showed a normal TSH response to TRH in six patients with KS. These contradicting reports indicate that the latent or subclinical hypothyroidism due to pituitary or hypothalamic defect, if present, is not universal in patients with KS.

Lingual thyroid is often associated with hypothyroidism (*11*). The cause of hypothyroidism in the patient presented is unknown. Since he had no evidence of mental retardation or retarded bone growth, the hypothyroidism was, most likely, an acquired one and was due to autoimmune thyroiditis as seen in other gonadal dysgeneses (*12*). However, the incidence of autoimmunity in KS has been found to be higher than, lower than, or the same as that in appropriate control groups (*13*).

KS and sublingual thyroid are both congenital abnormalities. Individually the conditions are rare, the incidence of KS being 1 in 1,000 phenotypic boys (*9*) and that of ectopic thyroid 1 in 100,000 general patients (*11*). In the case described here, these two rare entities are combined. A case with KS, congenital myxedema, and lingual thyroid has been described (*4*).

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