CALIFORNIA CANCER COMMISSION

SEMI-ANNUAL SLIDE CONFERENCE

ON

CHEMICAL AND HISTOLOGICAL CORRELATION OF SOME OF THE ENDOCRINE ABNORMALITIES

MODERATOR:

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CHAIRMAN:

CARL M. MC CANDLESS, M. D.
SAN FRANCISCO, CALIFORNIA

SUNDAY, DECEMBER 3, 1961

9:00 A.M. - 4:30 P.M.

MARK HOPKINS HOTEL
San Francisco, California

Please send in your diagnoses, using the separate sheet enclosed, on or before November 22, 1961, so that they may be tabulated before the meeting.

Please bring your protocol, but do not bring slides or microscopes to the meeting.

This conference for the most part has been chosen for the clinico-laboratory findings and not for the histopathology of the lesional tissue giving rise to the chemical and hormonal aberrations.

DIAGNOSES FOR CALIFORNIA CANCER COMMISSION SEMI-ANNUAL SLIDE CONFERENCE ON CHEMICAL AND HISTOLOGICAL CORRELATION OF SOME OF THE ENDOCRINE ABNORMALITIES. MODERATOR: GERSON R. BISKIND, M. D., Dec. 3, 1961

1. Testicular Feminizing Syndrome and Male Pseudohermaphroditism with Hyperplasia of Interstitial Tissue of Testicles
2. Leydig Cell Tumor of the Ovary
3. Granulosa Cell Tumor of the Ovary
4. Adeno-Lipoma of Parathyroid Gland
5. Choriocarcinoma in Term Placenta
6. Thymoma with Myasthenia Gravis
7. Adrenal Cortical Carcinoma with Adreno-Genital Syndrome
8. Bilateral Interstitial Cell Tumors in Salt-Losing Variant of Adreno-Genital Syndrome
9. Adenomatoid Hyperplasia of the Adrenal Cortex with Hypertensive Variant of Adreno-Genital Syndrome.
10. Islet-Cell Tumor of Pancreas with Hyperparathyroidism
11. Adrenal Cortical Adenoma with Aldosteronism
12. Phaeochromocytoma of Adrenal
13. Lung Tumor (Paraganglioma) with Cushing's Syndrome
14. Hydatidiform Mole
15. Oat Cell Carcinoma of Lung with Cushing's Syndrome
16. Choriocarcinoma of Testicle
17. Thecoma of the Ovary
18. Mesothelioma of Pleura with Hypoglycemia
19. Tubular Adenoma of the Ovary
20. Carcinoma of the Adrenal with Adreno-Genital Syndrome
21. Choriocarcinoma in Uterus
22. Bilateral Adrenal Hyperplasia with Adreno-Genital Syndrome, Salt-Losing variant, and Female Pseudohermaphroditism
23. Adrenal Cortical Adenoma with Cushing's Syndrome
24. Normal Adrenal Gland in Cushing's Syndrome
25. Embryonal Carcinoma with Suggestive Chorionic Elements in Testicle

CASE NO. 1 DECEMBER 3, 1961

ACCESSION NO. 10857 OUTSIDE NO. PSH 56-1186
MHG 57-101

NAME: M. N.
AGE: 24 SEX: Apparently female RACE: Caucasian

CONTRIBUTOR: Herbert I. Harder
Glendale Sanitarium & Hospital
Glendale, California

TISSUE FROM: Gonads

CLINICAL ABSTRACT:

History: These identical twins were reared as girls and were thought to be normal until puberty. They looked alike, had the same body build, and had the same clinical, anatomic, and histologic findings. They had never menstruated. Neither of them had pubic or vulvar hair and had only scant axillary hair. The breasts and nipples enlarged at times but there was never any periodic soreness of the breasts. They had occasional episodes of mild right lower quadrant pain described as coming and going for one to two days.

Both reported normal libido, dated, and had "normal feelings," enjoyed kissing, etc. They were more embarrassed by the absence of the pubic hair and less concerned by the absence of the vagina and uterus. They both worked in an office and later became airline stewardesses.

Physical examination: The heights were 5'5" and 5'5½"; weights, 115 and 116 pounds. Both had normal female body contour, normal breast development, but infantile nipples and pale pink areolae. There was scant axillary hair and only fuzz on the pubis and vulva. There were right inguinal hernial weaknesses but no masses or tenderness. The clitoris in each was small, the labia small, pink, infantile, and without the usual pigmentation. The vagina was 2 cm. in diameter and 5 cm. deep, and no cervix or uterus could be palpated. A mass, 4 cm. in diameter, was felt in each adnexa.

Course: They were treated with oral premarin and premarin vaginal cream. It was advised to delay the vaginal plastic surgery until a few months before marriage. Psychologically they were impressed that they would be able to live happy lives, fulfill marital obligations, but would never menstruate nor have children.

SURGERY:

On the first twin, exploratory surgery was performed in May 1956. The rudimentary gonads were found at the inguinal rings bilaterally. A rudimentary muscular structure, 3 x 3.5 cm. in diameter, presumably a uterus, was found on the medial aspect of each gonad. Extending laterally for 3 cm. from the lateral aspect of the rudimentary uteri were single rudimentary tubes, one of which had a cyst, measuring 1.5 cm. Across the bladder vault between the inguinal rings was a fibrous, narrow "round ligament." The urinary system appeared normal.

ACCESSION NO. 10857

Bilateral excisions of the gonads, rudimentary uteri and tubes were performed, as well as herniae repair and plastic vaginal procedure. The appendix was also removed.

The second twin was operated in January 1957. The findings were exactly as in twin No. 1 and the same structures were removed, the herniae repaired, and the vaginal plastic procedure done.

GROSS PATHOLOGY: (First twin)

The left and right adnexa were separately submitted and each included a rudimentary uterus partially fused to a gonad; the left one had a long tubular structure, measuring 3 cm. The left gonad was firm, 3.8 x 2.5 x 2.3 cm., and was mottled, pale, pink-gray-white. On section, it had a bright yellow-orange-tan parenchyma compressed to the periphery by a homogeneous, pink-gray, firm nodule, 1.9 cm. in diameter. The rudimentary uterus measured 3.2 x 2.0 x 1.5 cm. The gross specimen from the second twin was similar in size and appearance.

LABORATORY REPORT:

Urinary gonadotrophins were preoperatively and postoperatively less than 6 mouse units (Normal: 6 - 50 for both male and female).

Estrogens were preoperatively 17.4 ug/24 hr. and postoperatively 8.4 ug/24 hr. (Normal: 2 - 29 for 24 year old male; 4 - 48 for 24 year old female).

Urinary 17-ketosteroids were preoperatively 5.1 mg/24 hr. and postoperatively 4.6 mg/24 hr. (Normal: 11 - 27 for 24 year old male; 7 - 14 for 24 year old female).

Skin biopsy showed almost complete absence of the peripherally located intranuclear "sex chromatin" mass seen in the somatic cells of the female.

Blood smear showed a rare "drumstick" sex chromatin mass, but in numbers too small for the female sex.

FOLLOW-UP:

Both patients are "happy and eager and have an entirely natural outlook."

Reference: Marshall, H. K. and Harder, H. I. Testicular Feminizing Syndrome in Male Pseudohermaphrodite, Report of two cases in identical twins. Obst. & Gynec. 12:284-293, Sept. 1958.

CASE NO. 2 DECEMBER 3, 1961

ACCESSION NO. 10773 OUTSIDE NO. HM 2153-59

NAME: A. V.
AGE: 78 SEX: Female RACE: Unknown

CONTRIBUTOR: Lillian M. Rowan
Hoag Memorial Hospital
Newport Beach, California

TISSUE FROM: Right ovary

CLINICAL ABSTRACT:

History: The patient had been noted to have marked hirsutism in recent months, a prominent mustache, and hoarseness of the voice.

Physical examination disclosed a pelvic mass.

LABORATORY REPORT:

Urinary 17-ketosteroids were 145 mg/24 hr. (Normal: 6 - 15; Norymberski method). Urinary 17-ketogenic steroids totalled 68 mg/24 hr. (Normal: 4 - 15; Norymberski method).

SURGERY:

On October 12, 1959, a large ovarian mass was found to be densely adherent to the posterior serosal surface of the uterus, but did not appear to invade the uterine wall. The uterus and the ovarian mass were removed.

GROSS PATHOLOGY:

The ovarian mass measured 15 x 3 x 8 cm. On section, it was rather fleshy, pink-gray in color, with numerous areas of necrosis and pseudocystic degeneration. The uterus was enlarged for the patient's age (12 x 7 cm.), and the opposite ovary was atrophic, being 2 cm. in greatest dimension.

COURSE:

Six days after surgery, the 17-ketosteroids were 10 mg/24 hr. and six weeks after surgery, 4.9 mg/24 hr.

FOLLOW-UP:

The patient was seen by her family physician 8 months post-operatively and at that time there was no evidence of masculinization. The patient died from an apparent cerebrovascular accident in 1960. No autopsy was performed.

CASE NO. 3 DECEMBER 3, 1961

ACCESSION NO. 10305 OUTSIDE NO. S3128-58

NAME: E. C. R.
AGE: 65 SEX: Female RACE: Caucasian

CONTRIBUTOR: Maria I. Barrows
St. Joseph's Hospital
San Francisco, California

TISSUE FROM: Left ovary

CLINICAL ABSTRACT:

History: The patient was gravida III, para II, and 15 to 20 years postmenopausal. She had had vaginal spotting for four months and one episode of heavy bleeding 12 days prior to entry. She had no pain, cramps, or systemic symptoms.

SURGERY:

On May 7, 1958, a dilatation and curettage was performed. Only minute endocervical fragments were obtained which were not diagnostic. The uterus, tubes, and ovaries were removed on November 30, 1958.

GROSS PATHOLOGY:

The left ovary measured 3.5 x 2.5 x 2 cm. and on section revealed a sharply circumscribed, bright yellow tumor, 1.8 cm. in diameter. Grossly, this ovarian neoplasm resembled an adrenal cortical adenoma. Several corpora albicantia were visualized in the soft tissue peripheral to the tumor. The fallopian tubes, the right ovary, and the uterus were not remarkable. The cervix revealed multiple cysts, varying from 0.1 to 0.3 cm. in diameter.

FOLLOW-UP:

The patient has been in good health since the surgery, with the exception of one episode of jaundice in 1959 which was clinically diagnosed as hepatitis.

CASE NO. 4 DECEMBER 3, 1961

ACCESSION NO. 11463 OUTSIDE NO. S60-6996

NAME: N. R. S.
AGE: 43 SEX: Male RACE: Caucasian

CONTRIBUTOR: S. K. Abul-Haj
Walter Reed General Hospital
Washington, D. C.

TISSUE FROM: Retropharyngeal mass, right side.

CLINICAL ABSTRACT:

History: This 43 year old man had been hospitalized on November 13, 1960, because of severe right flank pain which subsided spontaneously after admission. Excretory urogram at that time revealed a right mid-ureteral calculus and a left renal pelvic calculus. Patient was discharged, but nine days later was admitted to Walter Reed General Hospital with the same complaint.

Past history revealed that sixteen years previously, an episode of dull flank pain occurred, lasting several days, and terminating with the passage of a calculus. Other than an episode of "mild cystitis" in 1958, he had been free of symptoms until the onset of present complaint.

Systemic review established that he had been aware for sometime of an "unusual non-descript sensation" in the right side of his throat.

Physical examination on admission revealed no unusual findings. Subsequent laboratory studies established a persistent elevated serum calcium. Almost three weeks after admission, a 3 x 4 mm. urinary tract calculus was passed.

Laboratory report: Routine hemograms and urinalysis were within normal limits. Serum calcium was 12.6 mgm%; alkaline phosphatase, 16.9 K.A.U.; and phosphorus, 2.5 mgm%. A phosphate tubular clearance was 30.7 cc/min. (Normal: Less than 15.0). This was not significantly reduced following parenteral infusion of 10 mg. Ca/Kg. body weight (820 mg. total Ca).

A radiologic bone survey of the skull, hands, feet, clavicles, and ribs revealed no lesions.

SURGERY:

On December 21, 1960, a neck exploration was done. A discrete, oval, smooth, retropharyngeal right-sided mass which extended inferiorly into the superior mediastinum was dissected free from the surrounding structures and removed. It had a vascular pedicle by which (continued on next page)

it was attached to the right inferior thyroid artery and vein. It was grossly encapsulated and clearly separate from the thyroid gland. No normal parathyroid glands were identified.

GROSS PATHOLOGY:

The specimen consisted of an ovoid, apparently encapsulated, smooth, rubbery firm mass, 5 x 3 x 2 cm., weighing 19.0 gm. The cut surface was oily to touch, yellow-tan, with lighter foci of opalescence, and irregular speckling by more solid green to orange areas.

LABORATORY SUMMARY:

	12-3-60	12-6-60	12-7-60	12-8-60
Urine vol. cc/24 hr.	-	3750.0	3985.0	3000.0
Total Ca excreted mgm/24 hr.	-	911.0	739	740
* Serum Ca mgm%	13.9	14.2	14.0	15.0
* Serum phosph. mgm%	2.0	1.4	2.2	2.0
Sulkowitch test	4+	4+	4+	-
Serum alk. phosph. (K.A.U.)	-	15.0	15.6	16.9

	12-23-60	1-18-61	1-20-61	1-22-61
Urine vol. cc/24 hr.	-	2390.0	2180.0	2600
Total Ca excreted mgm/24 hr.	-	13.0	66.0	9.0
* Serum Ca mgm%	9.8	10.4	10.2	11.0
* Serum phosph. mgm%	3.0	3.3	3.6	3.7
Sulkowitch test	-	Neg.	Neg.	-
Serum alk. phosph. (K.A.U.)	-	-	-	-

(* Normal range: Serum Ca, 9 - 11; serum phosph., 3.0 - 4.5)

COURSE:

Post-operative course was uneventful. Calcium gluconate was given for three days whenever necessary for positive Chvostek sign. He was discharged on the third day and had an uneventful recovery except for a transient episode of tetany on the sixteeth post-operative day which was treated with calcium. He was free of symptoms one month later.

FOLLOW-UP:

Routine check-up in September 1961 revealed no unusual findings and patient was free of symptoms.

CASE NO. 5 DECEMBER 3, 1961

ACCESSION NO. 11343 OUTSIDE NO. SF59-8224

NAME: J. B.
AGE: 44 SEX: Female RACE: Caucasian

CONTRIBUTOR: Milton L. Bassis
Permanente Medical Group
San Francisco, California

TISSUE FROM: Placenta

CLINICAL ABSTRACT:

History: At the time of her first admission to the hospital on December 5, 1959, the patient was gravida III, para I, abortion I, and her expected date of confinement was December 24, 1959. One month prior to admission, she noted a hardness of the abdomen with a gradual increase in size, cramps, dizziness, and dyspnea. Her blood pressure was 110/62, pulse 86, and respirations 28. Her abdomen was distended and the fundus of the uterus could be palpated at the xiphoid process. The fetal heart tones were heard in the lower right quadrant, 130 per minute. The cervix was tightly closed. She remained in the hospital two days for observation, during which time dyspnea increased and she developed pedal and pretibial edema. An evaluation of her cardiovascular system indicated it to be normal. It was thought that there might be excess fluid in the uterus and the fetal parts could not be identified. X-ray indicated a possible anencephalic monster.

Shortly after discharge, she returned because of increasing edema. The fetal heart tones could not be heard. Blood pressure was 120/78, pulse 90, and respirations 16. The fundus was 3 cm. below the xiphoid process; the uterus was distended with fluid, without fetal movement, fetal heart tones, or contractions.

On December 14, 1959, a stillborn anencephalic female infant was delivered spontaneously An autopsy of the infant disclosed a typical anencephalic monster with extensive maceration and autolysis of all organs; no evidence of tumor was found.

The following day, the patient developed fever, the x-ray showed some infiltration in the right lower lobe, probably an aspiration pneumonitis (patient was thought to have aspirated fluid during the delivery). This cleared gradually with antibiotics and supportive measures.

ACCESSION NO. 11343

Laboratory report: On December 18, 1959, a serum pregnancy test (frog) was performed which was reported as positive. On December 20, 1959, the serum pregnancy test (frog) was positive in a dilution of 1:100. On December 29, 1959, the qualitative pregnancy test was positive, although the uterus was fairly well involuted but somewhat boggy.

Chest x-rays were again taken and compared with the ones taken earlier and the report indicated that metastatic disease should be considered in addition to the pneumonitis.

SURGERY:

A total hysterectomy and left salpingo-oophorectomy was performed on December 31, 1959, and a biopsy of a tumor nodule on the lower anterior vaginal wall was accomplished. This tumor nodule had not been present at the time of delivery.

GROSS PATHOLOGY:

The placenta measured 18.5 x 17 cm. in greatest dimensions; the centrally inserted umbilical cord measured 29.5 cm. in length. The cotyledons were intact and presented a coarsely granular, very friable, pink-yellow appearance. Many solid, gray-yellow, sharply demarcated tumor nodules were seen to replace about two-thirds of the normal placenta.

COURSE:

A quantitative serum frog pregnancy test was done on January 6, 1960 which was reported as positive undiluted, but negative in dilution of 1:10. The patient was started on methotrexate therapy. The chest x-ray presented no significant change in the infiltrated areas and it was considered that the patient had metastatic disease in the lungs. The gonadotrophin titer was positive 1:100, but negative 1:1000.

On January 26, 1960, four purple nodules on the anterior vaginal wall near the introitus were seen, one of which bled easily. On February 29, 1960, the serum pregnancy test was negative and was again negative on April 18, 1960. The patient improved; the vaginal nodules regressed. On July 3, 1960, the urinary gonadotrophins/24 hr. were reported positive at 1000 mouse units, negative at 5000 mouse units.

X-rays in June 1960 revealed metastatic nodules in the right lower lobe, and the gonadotrophin titer was positive at 1000 mouse units/24 hr.

Repeat chorionic gonadotrophin titer in November 1960 was negative at 500 mouse units, and the x-ray of the chest indicated a marked regression of the lesions as compared with the previous films. The patient had been treated with nitrogen mustard in September because of the positive pregnancy test.

ACCESSION NO. 11343

FOLLOW-UP:

Quantitative Aschheim-Zondek tests were performed by the standard technique on mice and from January through July 1961 were negative at 500 mouse units. In August 1961, the test was positive at 1,000 mouse units, but negative at 5,000. By the modified Reddy extraction method, the 17 hydroxy-corticoids were 6.9 mg/24 hr. (Normal: 4 - 10) in July 1961.

The patient received two more courses of methotrexate, 5 mg. b.i.d. for 3 days, the first in February and the second in April of 1961. The white blood cell and platelet count remained normal. Following the rise in gonadotrophins in August, another course of methotrexate was given, this time 10 mg. q.i.d. for 4 days.

Chest x-rays in April 1961 showed a slight increase in the pleural effusion at the left base, which now obscured the left diaphragm and costophr angle. There was no change in the size of a 1.5 x 3 cm. nodule in the right second interspace anteriorly. A poorly-defined 1 cm. nodule was present in the right lower lung field. No evidence of bony metastasis was seen. Repeat films on July 25, 1961 showed the pleural effusion to be absent and no new nodules were seen.

Follow-up received on October 30, 1961, states that the patient is alive, and except for mild symptoms referable to the chest associated with a mild intermittent cough, appears well.

CASE NO. 6 DECEMBER 3, 1961

ACCESSION NO. 10650 OUTSIDE NO. S-4852-59

NAME: J. F.
AGE: 48 SEX: Male RACE: Unknown

CONTRIBUTOR: George Hummer
St. John's Hospital
Santa Monica, California

TISSUE FROM: Anterior mediastinal mass

CLINICAL ABSTRACT:

History: The patient complained occasionally of double vision and ptosis of the left eye. Clinical evaluation in June 1956 led to the diagnosis of myasthenia gravis.

The patient was re-admitted in October 1959 because of the recurrence of ptosis. The angiogram was essentially normal. X-ray of the chest revealed a tumor of the anterior mediastinum.

SURGERY:

On October 22, 1959, an exploration of the anterior mediastinum was done and a firm mass, 6 cm. in diameter, was found lying to the left of the sternal space at about the level of the bifurcation of the pulmonary artery. By sharp and blunt dissection, the encapsulated mass was excised.

GROSS PATHOLOGY:

The specimen consisted of an oval firm tumor mass, measuring 8 x 7 x 5.5 cm. and weighing 130 gm. The surface was coarsely nodular. On section, it was composed of fleshy, pink-tan stroma separated by thin fibrous septae.

FOLLOW-UP:

Patient was last seen on September 7, 1961, at which time he had slight lid ptosis.

CASE NO. 7 DECEMBER 3, 1961

ACCESSION NO. 10724 OUTSIDE NO. C-1340-59

NAME: P. A. E.
AGE: 24 SEX: Female RACE: Caucasian

CONTRIBUTOR: Wayne Scott
Community Hospital
San Bernardino, California

TISSUE FROM: Right adrenal

CLINICAL ABSTRACT:

History: This patient was first seen by her physician on March 30, 1959, with a history of having increasing facial hair, development of male pattern pubic hair, and growth of sternal hair over the past twelve months. The breasts were decreased in size and pendulous, and the menses had been irregular for six months. She had been told at a previous examination that she had an ovary about the size of a walnut on the right. At this time the ovary was considerably larger and the uterus was normal in size. Progesterone therapy was given and a menstrual flow (normal) occurred. On May 29, 1959, she returned to the office, twenty-two days past due, and stated that facial hair had increased in amount. Urinary 17-ketosteroids at this time were 190 mg/24 hr. (Normal by method used: 5 - 14 mg/24 hr). This was fractionated; A fraction was 108 mg. and B fraction was 82 mg. Pneumography revealed a large mass in the right adrenal area.

Past history revealed that menarche occurred at the age of 12 and menses were fairly regular and normal in flow but accompanied by severe dysmenorrhea. The dysmenorrhea disappeared following first delivery. Patient is para II, gravida, II, both normal vaginal deliveries.

Physical examination: The increased facial hair, male escutcheon, sternal and areolar hair, and atrophic pendulous breasts were confirmed. A suggestion of a mass or increased resistance in the right upper quadrant of the abdomen was also found.

SURGERY:

On June 13, 1959, exploration of the right upper quadrant mass revealed a yellow, soft, globular tumor, 11 cm. in diameter, replacing the adrenal gland. It was adherent to the diaphragm but separate from the kidney.

GROSS PATHOLOGY:

The specimen consisted of a yellow, soft, roughly globular tumor, 11 cm. in diameter. The cut surface bulged and was mottled with pinkish gray to tan soft areas and yellow, firmer friable streaks.

ACCESSION NO. 10724

COURSE:

Almost the entire mustache, beard, and sternal hair disappeared. The male escutcheon remained but was much reduced. Patient became pregnant and delivered a normal male infant on August 24, 1960. In September 1960, chest x-ray revealed densities suggestive of metastatic nodules.

FOLLOW-UP:

The patient was referred to the UCLA Medical Center where a biopsy of a recurrent right upper quadrant abdominal mass was done. The biopsy report described a tumor similar to the original surgically resected mass. On October 11, 1960, a 17-ketosteroid was done by the Norymberski method and found to be 52.3 mg/24 hr. (Normal: 5 - 20). A 17-hydroxycorticosterone was done by the Porter and Silber method and was 9.4 mg/24 hr. (Normal: 2.9 - 10.3). The patient began losing weight and symptoms of urinary tract infection developed. Pyelography revealed obstructive uropathy secondary to impingement of the outflow tract of the right kidney by the tumor mass. Repeat chest film showed increase in size of the pulmonary mass. On December 12, 1960, the patient was started on D.D.D. (an inhibitor of adrenal hormone synthesis). On this medication, the mass diminished in size and steroid studies done on January 19, 1960 were as follows: 17-ketosteroids were 3.7 mg/24 hr. and 17-hydroxysterone was 7.7 mg/24 hr. (Normal: 5 - 18). The patient has continued taking D.D.D. and when seen in September of 1961, the abdominal mass was not palpable and no lesions were seen on the chest film. Except for occasional nausea and vomiting she has felt well. There has been a return of normal regular menses.

CASE NO. 8 DECEMBER 3, 1961

ACCESSION NO. 10825 OUTSIDE NO. S59-3993

NAME: J. M.
AGE: 16 SEX: Male RACE: Caucasian

CONTRIBUTOR: N. L. Morgenstern
Kaiser Foundation Hospital
Oakland, California

TISSUE FROM: Right testis.

CLINICAL ABSTRACT:

History: The patient was born on April 22, 1943, after an uneventful pregnancy. Birth weight was 3410 grams. The siblings were normal. At the age of two weeks, he had projectile vomiting and was treated by hypodermoclysis and supportive therapy. He was transferred to the University of California Hospital at the age of 2½ months, weighing only 3060 gm. and markedly dehydrated. The penis was noted to be large. The serum sodium and chloride were low, and the diagnosis of Addison's disease was made. He improved on treatment with adrenal cortex extract (Eschatin), intravenous and oral salt therapy. The weight was not maintained until the dose of adrenal cortex extract had been increased to 24 cc. daily and desoxycorticosterone acetate was added. The dose was gradually reduced and finally omitted entirely, and the child was discharged from the hospital; his serum sodium was 134 mEq/L. He was treated at home by sublingual desoxycorticosterone acetate and oral salt.

During the second year, the patient showed advanced bone age and precocious sexual development. At the age of 16 months, the urinary 17-ketosteroids were 5.9 mg/day, and the bone age , which had been equal to his chronological age at 11 months, was advanced to that of a normal 30 month old child. IQ was 106.

A trial period without adrenal therapy was attempted, and he lost 600 gm. in weight within three days, began to vomit, and became dehydrated. Resumption of therapy promptly relieved the symptoms. At the age of 2½ years, pubic hair appeared. The urinary 17-ketosteroid level was 14.9 mg/24 hr. When he was 3 years old, his bone age was 10 years. At the age of 4 years he was 44 inches (112 cm.) tall, and acne was present. Six months later he had a deep voice, the penis was 10 cm. long, and the testes were the size of those of a prepubertal youth. At this time a pellet of desoxycor ticosterone was implanted in the skin.

At the age of 6 years and 2 months, he was 52 inches tall, average for 9 years of age. No abnormal pigmentation of the skin was present, and the dentition appeared normal. At the age of 7 years and 3 months, he weighe 80 lbs. (36 kg.), and was 54 inchest (137 cm.) tall. He would occasionally take sodium chloride in addition to what was placed in his food. At this time he had a slight amount of axillary hair, slight acne, and a progressivel deepening voice. The growth in height ceased at the age of 8 years.

When the patient was 10 years old, biopsy of both testicles revealed normal prepubertal testes with no evidence of spermatogenesis and normal-appearing Leydig cells. Cortisone therapy was begun (25 mg. t.i.d.), but the medication was not taken regularly. When he was 14 years old, the size of the testes appeared normal for an adolescent male. He had some emotional problems which were attributed to his short stature. On two occasions during 1957, urinary 17-ketosteroid excretions were 12.6 and 21.6 mg/24 hr. The blood pressure was consistently normal.

The patient was seen at the Endocrine Clinic of Kaiser Foundation Hospital, Oakland, on November 13, 1957 at the age of 14 years, seven months after a mild Addisonian crisis. He was short and very muscular with small hands and feet. The penis was large and the testes appeared normal in size and consistency for his age. Blood pressure was 92/64. The urinary 17-ketosteroid level was 16.2 mg/24 hr. and the 17-hydroxycorticosteroid level was 7.9 mg/24 hr.

The patient did not return until one year later in November 1958. The diameter of each testis was now about 35 cm., and they were hard. The 24-hour urinary 17-ketosteroid value had increased to 90 mg. and the 17-hydroxycorticosteroid level had diminished to 0.2 mg/24 hr. Treatment with cortisone was again begun, and it was now taken regularly. On March 27, 1959, the 17-hydroxycorticosteroid excretion had risen to 8.2 mg/day, but the 17-ketosteroid level was 142 mg. One month later the corresponding values were 18.6 mg. and 95.6 mg/day.

SURGERY:

On June 19, 1959, bilateral testicular biopsies and a right orchiectomy were performed. The left testis increased in size and on September 29, 1959, a left orchiectomy was performed. Plastic prosthetic testes were placed in the scrotum.

GROSS PATHOLOGY:

The right testis measured 5 x 2 x 2 cm. and weighed 55 gm. The testicular parenchyma was replaced by tumor except for a very thin shell along one margin. The tumor was quite firm with brownish-gray trabeculated sectioned surfaces. (The left testis measured 3 x 4.5 x 4 cm. and weighed 100 gm. The tumor grossly resembled that of the right testis.)

SPECIAL LABORATORY TESTS:

Effect of ACTH stimulation: Administration of 25 units of ACTH in 1000 ml. five percent dextrose in water, given over an 8-hour period, done prior to orchiectomy produced no significant increase in the urinary 17-ketosteroid excretion. Hydroxycorticosteroid level increased from 18.6 to 32.8 mg/day.

ACCESSION NO. 10825

Following right orchiectomy, urinary 17-ketosteroid values rose from control levels of 38.6 and 43.1 mg. to 75.7 and 79.5 mg. on administration of ACTH on two separate days. No apparent change in the level of the urinary 17-hydroxycorticosteroids was noted. After the second orchiectomy, 17-ketosteroid excretion values showed no rise and 17-hydroxycorticosteroid was not altered.

Prior to orchiectomy, there was a marked fall in the 17-ketosteroid values when fluorohydrocortisone was administered in addition to cortisone. Unilateral orchiectomy resulted in a fall of the urinary 17-ketosteroid level to about half, and bilateral orchiectomies to approximately normal.

On September 1, 1959, while the patient was receiving fluorohydrocortisone, 8 mg. daily, 500 units of chorionic gonadotrophin were administered intramuscularly for 3 days. On the first day, the ketosteroid values doubled, but on the second and third days fell to levels near those observed prior to administration. The 17-hydroxycorticosteroid excretion was not noticeably affected.

Pregnanetriol excretion: 66 mg/day prior to removal of the tumors.

Urinary gonadotrophin: Urinary gonadotrophin values were less than 5 mouse units prior to orchiectomy and after the right orchiectomy.

In November 1959, one month following bilateral orchiectomies, the urinary gonadotrophins had risen to more than 5 mouse units and less than 80 mouse units daily.

Sex chromatin pattern was male.

COURSE:

He did well post-operatively and has been maintained on fluorohydrocortisone (0.5 mg. t.i.d.). The urinary 17-ketosteroids and the 17-hydroxycorticosteroid excretion values were similar to those observed in 1957.

FOLLOW-UP:

A month after surgery, a hematoma developed requiring drainage. Shortly thereafter, because of infection, replacement of prosthesis was required. In April 1960, 17-ketosteroids were 19.1 mg/24 hr. He has been seen recently (1961) with no apparent change in status.

References: Schoen, E., DiRaimondo, V., and Dominguez, O. V. Bilateral testicular tumors complicating congenital adrenocortical hyperplasia. J. Clin. Endocrin. & Metab., 21:518-532, May 1961.

Deamer, W. C., and Silver, H. K. Abnormalities in the secretion of the adrenal cortex during early life. J. Pediatrics, 37:490, 1950 (Case No. 3).

CASE NO. 9 DECEMBER 3, 1961

ACCESSION NO. 11383 OUTSIDE NO. 398-61

NAME: H. R.
AGE: 46 SEX: Male RACE: Unknown

CONTRIBUTOR: D. A. DeSanto
Mercy Hospital
San Diego, California

TISSUE FROM: Left adrenal

CLINICAL ABSTRACT:

History: This patient was admitted to the hospital on January 10, 1961, complaining of swollen legs of three weeks' duration. He is a known hypertensive and had been on intermittent therapy since 1943. In 1952, he was admitted for a hemorrhoidectomy and at that time his blood pressure was 155/95. Until the present symptoms, he had been doing well except for marked anxiety. He had previously been free of dyspnea, orthopnea, headaches, dizzy spells, and only recently developed nocturia x 3 and gained 15 lb. weight. His mother, age 78, is a diabetic.

Physical examination: Patient was a well developed, well nourished, slightly obese male with a blood pressure of 210/115. Four plus edema was present to the mid-calves. Cardiac border of dullness extended to the anterior axillary line. Circulation time (arm to tongue) was shortened and the venous pressure was elevated

Laboratory report: Admitting urinalysis had a specific gravity of 1.012 and a 3 plus sugar reaction. Fasting blood sugar was 134 mg% and postprandial blood sugar was 308 mg%. Other studies were as follows: BUN 14.5 mg%; CO_2 90 vol% (50-70); HCO_3 40 mEq/L (22-33); Cl 86 mEq/L; Na 150 mEq/L; K 2.7 mEq/L; BSP 25%; blood cholesterol 304 mg%. Urinary 17-ketosteroids on January 17, 1961 were 42 mg/24 hr. (Normal: 9 - 22); 17-ketogenic steroids, 43 mg/24 hr. (Normal: 8 - 25). Urinary 17-ketosteroids on January 23, 1961 totalled 49 mg/24 hr.; urinary 17-hydroxie steroids (Glen Nelson method) 15.4 mg/24 hr. (Normal: 3 - 10). Urinary ketosteroids on January 24, 1961 were 64 mg/24 hr; 17-ketogenic steroids (Norymbersky method), 68 mg/24 hr.

X-ray report: Excretory urograms were negative, however, nephrotomograms were suggestive of a mass in the region of the left adrenal. Chest x-rays showed left ventricular hypertrophy.

SURGERY:

An exploratory laparotomy was performed on January 25, 1961. A large abscess-like mass of the body and tail of the pancreas was found which proved to be a subacute pancreatitis. This was removed along with the body

and tail of the pancreas and the spleen, following which the left adrenal was explored and found to be enlarged and was removed. At this time the right adrenal gland was palpated and seen to be equally enlarged, but it was not removed because of the lengthy operation. Following surgery, the patient's blood pressure was 160/100 and again 160/90. The patient had various bouts of diaphoresis. He received insulin as needed.

GROSS PATHOLOGY:

The specimen was an adrenal, 6 x 4 x 1.5 cm. and weighed 16 gm. The cortex measured up to 5 mm. in thickness and was bright yellow. The thickening was uniform throughout the gland.

COURSE:

Multiple electrolyte studies were done and found to be within normal limits. The steroid studies were repeated post-operatively and were as follows: Urinary 17-ketosteroids on January 26, 1961 were 62 mg/24 hr; 17-hydroxie steroids, 17.8 mg/24 hr. Urinary sodium was 27 mEq/24 hr. vol. and potassium was 24.3 mEq/24 hr. vol. (January 30, 1961).

FOLLOW-UP:

Because of the finding of an area of pancreatic destruction, it was decided not to remove the opposite adrenal gland. Therefore, the patient was treated with irradiation to the pituitary body, the dosage and details are not available at this time. Following this course of treatment, the patient apparently made a fine recovery. His hypertension was well controlled and the patient seemed, to all intents, to be "cured."

CASE NO. 10 DECEMBER 3, 1961

ACCESSION NO. 10742 OUTSIDE NO. A 116-59

NAME: I. M.
AGE: 47 SEX: Female RACE: White

CONTRIBUTOR: John H. Rowe
Kaiser Foundation Hospital
Los Angeles, California

TISSUE FROM: Pancreas (autopsy)

CLINICAL ABSTRACT:

History: The patient had enjoyed good health all her life until three months prior to admission to the hospital. At that time she noted the onset of weakness, nausea, polydypsia, anorexia, and occasional vomiting. She was not aware of polyuria, although she had noted frequency of urination and some dysuria. She had also noted a change in her bowel habits with an increase in frequency from one to three times daily; her stools were also of very loose consistency but without gross pus or blood.

During this time she had lost 15 pounds of weight and had become aware of the increasing prominence of her upper abdomen. In association with the increased prominence of her upper abdomen, she complained of intermittent cramping pains of the lower abdomen, lasting from 15-30 seconds. She had also become amenorrheic. There was no history of fever, chills, or jaundice. Past history was negative except for the fact that her mother had died of an abdominal malignancy at the age of 45 years.

Physical examination revealed a 2 cm. nodule in the lower pole of the right lobe of the thyroid. The abdomen was protuberant and the liver was enlarged, the lower margin being palpated four finger breadths below the costal margin in the right midclavicular line. The inferior surface of the liver felt irregular and a 4 x 4 cm. nodule was palpated in the right lobe; a similar nodule could be felt in the left lobe.

Chest x-ray, barium enema, and intravenous pyelogram were normal. The gastrointestinal series was normal and the skull, spine, and long bone series revealed no evidence of metastatic tumor. The bones were moderately demineralized but not deformed.

Laboratory report: The alkaline phosphatase was 32.9 K-A units (normal 9-11); serum albumin 3.4 gm/100 ml. and globulin 4.0 gm/100 ml. Blood calcium (May 27, 1959) was 19.2 gm/100 ml. and the phosphorus 2.2 mg/100 ml. Serum chloride was 87 mEq/L, serum sodium 143 mEq/L, and serum potassium 2.6 mEq/L. The Sulkowitch test performed on the urine was interpreted as showing a moderate amount of calcium.

ACCESSION NO. 10742

SURGERY:

Emergency surgery on May 28, 1959 was performed and the neck was explored for a parathyroid adenoma. A right thyroidectomy was done, and the frozen section revealed a nodule in the right lobe to be a colloid nodule. The root of the neck, the paraesophageal and the superior portion of the mediastinal fat pad were explored, and two small parathyroid glands were removed, but no adenoma was found. Also a small incision was made over the left lobe of the liver and a biopsy was obtained. A mass was palpated inferior to the liver in the region of the pancreas.

COURSE:

During the immediate post-operative period the patient was semi-comatose, but on the second day she became alert. The serum calcium on the second post-operative day was 15.8 mg/100 ml. She was treated with hydrocortisone preoperatively and post-operatively. On the sixth post-operative day the serum calcium was 14.7 mg/100 ml., but on the eighth day it had again risen to 19.4 mg/100 ml. (The hydrocortisone had been discontinued on the seventh day to obtain a 24-hr. urine specimen for evaluation of her calcium and phosphorus metabolism). She became very lethargic, confused and had fecal incontinence. On resumption of large doses of hydrocortisone, the serum calcium dropped to 15.7 mg/100 ml. for two days, and the patient cleared mentally but the diarrhea continued.

Despite continued administration of cortisone, the serum calcium rose to 19.4 mg/100 ml. Sodium versonate was added to the regime, and the serum calcium dropped to 15.9 mg. The liver was noted to be increasing in size. When the patient was preterminal, the serum calcium rose to 22.4 mg/100 ml., and she expired on the twenty-third post-operative day.

At autopsy careful dissection of the mediastinum and neck revealed no evidence of tumor. A parathyroid gland, measuring 0.4 cm., was identified and removed from the posterior portion of the left lobe of the thyroid. A circumscribed, gray-white granular tumor, 6 cm. in diameter, was found in the body of the pancreas. Grossly, the tumor appeared to be surrounded by dense fibrous connective tissue. Metastatic tumor was found within the liver and parapancreatic lymph nodes. The primary tumor of the body of the pancreas grossly invaded the portal vein at the point where the splenic vein and the superior mesenteric vein joined. Multiple sections from the pelvis, vertebrae, and ribs revealed no evidence of metastatic tumor.

Bioassay of segments of the tumor from both the liver and the pancreas revealed a parathormone effect when administered to parathyroidectomized animals. This study was performed through the courtesy of Dr. C. Kleeman at the Los Angeles Veterans Administration Hospital.

CASE NO. 11 DECEMBER 3, 1961

ACCESSION NO. 11694 OUTSIDE NO. 59-12916

NAME: R. B.
AGE: 33 SEX: Male RACE: Caucasian

CONTRIBUTOR: Dorothy Tatter
Los Angeles County Hospital
Los Angeles, California

TISSUE FROM: Right adrenal

CLINICAL ABSTRACT:

History: The patient consulted a doctor in 1957 because of persistent right temporal headaches. At that time he was discovered to be hypertensive. Since that time he has had visual blurring, polydypsia, polyuria, and irritability. The hypertension (blood pressure 210/130) was treated with "pills thought to be tranquilizers," and seemed to help him.

Physical examination: On admission to the Los Angeles County Hospital on August 16, 1959, his blood pressure was 220/140 bilaterally; pulse 72, regular. He had a grade III retinopathy (tortuosity, arteriolar narrowing, A-V nicking, exudates and flame-shaped hemorrhages). The heart was not enlarged. There was a grade II, soft, blowing systolic murmur heard best in the primary aortic area, which transmitted to the left sternal border and to the apex. The rest of the physical examination was normal.

Laboratory report: Routine tests were normal. Urinary catechol amines (method of R. J. Henry and C. Sobel: A.M.A. Archives of Internal Medicine. 100: 196-200, August, 1957) 3 micrograms/100 ml. (normal up to 14 ug%); serum sodium 148 mEq/L; potassium 2.1 mEq/L; chloride 90 mEq/L; creatinine 1.4 mg/100 ml. (normal). Urinary excretion (12-hour specimen 2 P.M. to 2 A.M. total volume 565 ml) of sodium 4.4 mEq, potassium 18 mEq, chloride 15.4 mEq, preformed creatinine 516 mg, phosphorus 0.35 g., and calcium 81 mg. or 4.1 mEq. Exchangeable sodium 3535 mEq., 48.3 mEq/Kg. (normal 37.8-47.5); exchangeable potassium 2121 mEq., 28.9 mEq/Kg. (normal 37.8-62.3); urinary uropepsin 104 and 81 units/hr. (normal 15-40); urinary aldosterone 32 gamma/24 hr.

X-rays of the chest and abdomen, and intravenous urogram did not reveal any masses.

SURGERY:

On September 18, 1959, an exploration was performed. The left adrenal appeared normal. The right adrenal had a golden yellow nodule, measuring 2 cm. A right adrenalectomy and an incidental splenectomy were done.

GROSS PATHOLOGY:

The right adrenal was submitted in multiple fragments. The cortex measured 0.2 cm. in thickness. The nodule, which measured 2 x 1.5 x 1 cm. and weighed 2 grams, was yellow-orange, uniform, and symmetrical.

COURSE:

The patient became shocky and was taken back to surgery. About 4000 cc. of blood were found in the operative site, apparently coming from the right adrenal artery. The vessel was ligated. The patient did well and was discharged on October 8, 1959. Chemistries on October 6, 1959 were: Serum sodium 130 mEq/L.; potassium 5.0 mEq/L; chloride 91 mEq/L. Blood pressure was 150/110 (October 26, 1959).

FOLLOW-UP:

The patient was last seen in the Out-patient Department of the Highland Alameda County Hospital on October 5, 1960: "Blood pressure 180/130; weight 171 pounds. Has not taken medication for three weeks, but follows salt instructions." Patient was restarted on reserpine, diuril, apresoline and phenobarbital. Patient did not keep his next clinic appointment.

CASE NO. 12 DECEMBER 3, 1961

ACCESSION NO. 10390 OUTSIDE NO. 016-31-69

NAME: R. A.
AGE: 9 SEX: Male RACE: Caucasian

CONTRIBUTOR: Sidney C. Madden
University of California Medical Center
Los Angeles, California

TISSUE FROM: Left adrenal

CLINICAL ABSTRACT:

History: For two years the patient had had flushing spells, associated with vomiting and fever. Retroperitoneal air studies showed a right adrenal tumor, and a right pheochromocytoma was removed. For fifteen months the patient was asymptomatic. Nine months prior to admission, the spells returned. Blood pressure was 210/160. Catechol amines were 565 micrograms/100 ml. (Hingerty technique; normals up to 30 mcg%).

Family history revealed that the patient's father died after a 17 year history of hypertension and paroxysmal attacks of sweats and nausea. Autopsy showed bilateral adrenal tumors, type unknown.

SURGERY:

On October 17, 1958, the lesion was excised from the left adrenal.

GROSS PATHOLOGY:

The tumor was firm, orange, encapsulated, and measured 7 x 3.5 x 2.5 cm. and weighed 38 gm.

FOLLOW-UP:

The patient postoperatively has been asymptomatic for three years with normal blood pressures and repeatedly normal catechol amines. He has been maintained for the past year on cortisone only.

CASE NO. 13 DECEMBER 3, 1961

ACCESSION NO. 022 OUTSIDE NO. 49-9785

NAME: M. B.
AGE: 38 SEX: Female RACE: Caucasian

CONTRIBUTOR: Harold Fanselau
Los Angeles County Hospital
Los Angeles, California

TISSUE FROM: Mediastinal mass

CLINICAL ABSTRACT:

History: About October 1948, the patient noticed increasing weakness associated with a rapid heart rate and swelling of legs and feet. Her physician found that she was hypertensive and in failure. Digitalis was prescribed. Approximately three months later, there was a noticeable increase in body hair and the patient experienced "hot flashes" as well as numerous bouts of epistaxis. Darkening of the skin of the arms and face occurred, and small hemorrhagic spots were noted in these pigmented areas as well as on the trunk but not in the lower extremities. In July of 1949, glycosuria was found. Dietary restrictions and insulin were used to control it. Exertional dyspnea, "hot flashes," nervousness, and severe headaches increased and the patient was admitted to the hospital on September 2, 1949.

Physical examination on admission revealed a well developed, obese, somewhat dusky and plethoric lady with obvious dyspnea and orthopnea. A moon facies was evident as well as a prominent "buffalo" hump and glossy pigmentation of the face and shoulders. Generalized purpura, abdominal stria, and male hair distribution were present. Blood pressure of 186/105, a pulse of 130, and respirations of 40/min. were recorded. Visual fields and funduscopic examination were negative. Fine basal rales and cardiomegaly were found associated with pitting edema. Bilateral Babinski reflexes were elicited.

Routine laboratory work revealed the hemoglobin to be 13.5 gm. with 9,500 white count and a normal differential. Urine specific gravity was 1.027 with a trace of albumin and a three plus reaction for sugar. Fasting blood sugar was 275 mg% and NPN was 41 mg%. Serology was negative. Electrocardiogram showed only left axis deviation. On September 7, a two-hour glucose tolerance test was done. The values obtained were for the fasting, ½ hr., 1 hr., and 2 hr. periods; 80, 167, 278, and 374 mg% respectively. Urine sugars for the same time periods were zero, 1 plus, 2 plus, and 4 plus. Plasma sodium, potassium, and chloride were within the normal range; the PBI was 6.6 microgram%.

On September 22, 1949, the 17-ketosteroids were found to be 28.8 mg/24 hr. (Normal: 6 - 12). Urine volume was 2000 cc.

A chest x-ray revealed a rounded mass which measured approximately 5 cm. in greatest diameter and appeared to be in the posterior left chest behind the breast and immediately adjacent to the lateral border of the 9th and 10th vertebral bodies. Sella turcica films and retrograde pyelogram studies were negative.

SURGERY:

On October 11, 1949, a left thoracolaparotomy was done. A mass arising in the inferior pulmonary ligament and extending up into the pulmonary tissue of the left lower lobe was removed. The left adrenal was also visualized and biopsied through a diaphragmatic approach. It appeared grossly and microscopically normal. There was no marked fall in blood pressure following removal of the tumor. She was returned to the ward with normal pulse, blood pressure, and respirations. The blood pressure remained normal, and the patient's post-operative course was uneventful except for pneumonitis which responded rapidly to antibiotics.

GROSS PATHOLOGY:

The specimen consisted of an egg-shaped encapsulated tumor mass which measured 5.5 x 3 x 3 cm. and weighed 35 gm. Externally it was tan with several areas of dark blue. Cut surface was a glistening light tan with numerous small areas of hemorrhage.

COURSE:

The patient was discharged from the hospital on October 28, 1949. There was marked symptomatic improvement. In January of 1950, the patient was admitted for follow-up studies. The hirsutism and pigmentation had decreased and the blood pressure was 110/70. The patient was normal appearing with no symptoms. Menstruation had returned. Physical examination, except for a murmur suggestive of aortic stenosis, was unremarkable. Seventeen ketosteroids were now 4.4 mg/24 hr. Glucose tolerance test was now normal and there was no glycosuria.

FOLLOW-UP:

In December 1957, the only complaint was a tendency to gain weight easily. The patient was seen at the Los Angeles County Hospital, April 1959, in the Surgical Clinic complaining of left anterior chest pain. Chest x-ray was ordered which showed the soft tissues, bony thorax, diaphragms, and cardiomediastinal silhouette to be normal. There was an ill-defined density in the left first anterior interspace. Apical and lordotic views were requested, however the patient failed to return for follow-up.

CASE NO. 14 DECEMBER 3, 1961

ACCESSION NO. 11767 OUTSIDE NO. 61-6164

NAME: L. G.
AGE: 30 SEX: Female RACE: Caucasian

CONTRIBUTOR: Kenneth Falconer
Los Angeles County Hospital
Los Angeles, California

TISSUE FROM: Uterine cavity

CLINICAL ABSTRACT:

On February 5, 1960, this patient entered the hospital in shock due to severe vaginal bleeding and cramping. Her last menses was in September 1959. The patient had considered herself pregnant and had experienced morning sickness. The breasts had enlarged and were tender. On admission, the fundus was 5 cm. above the umbilicus and contractions were every 5-8 minutes. No signs of toxemia were noted. Hemoglobin was 5.5 gm., clotting time was 3 minutes. No fetal heart tones could be heard and shortly after admission that night 12,000-15,000 cc. of cystic, grape-like material was spontaneously passed.

COURSE:

2-5-60: Urine obtained on admission gave positive Friedman test. Dilatation and curettage - tissue diagnosis: hydatiform mole.

2-7-60: Chorionic studies reported greater than 200,000 but less than 400,000 IU/L.

2-8-60: Repeat dilatation and curettage - tissue diagnosis: hydatiform mole.

2-11-60: Discharged from hospital.

3-17-60: Re-entered the hospital for moderate vaginal bleeding.

3-20-60: Chorionic gonadotrophins reported as greater than 10,000 but less than 40,000 IU/L.

3-21-60: Repeat dilatation and curettage - tissue diagnosis: hydatiform mole.

4-5-60: Chorionic gonadotrophins greater than 10,000 but less than 40,000 IU/L.

5-10-60: Friedman test reported as negative.

8-2-60: Friedman test reported as negative.

9-23-60: Because there had been no menses since last normal period in September 1959, a dilatation and curettage was done. Practically no tissue was obtained; reported as quantity not sufficient for diagnosis.

10-17-60: Chorionic gonadotrophins reported as less than 1,000 IU/L.
Pituitary gonadotrophins (FSH & LH) were reported as greater than 6 but less than 16 M.U.U. (normal 6-50 M.U.U.).

5-4-61: Protein bound iodine 5.7%. Cholesterol 206 mg.%.

5-9-61: Pregnandiol 3.8 mg/510 cc (24 hr. specimen).
Estrogen fractions: Estrone 2 ugm.
Estradiol 3 ugm.
Estriol 5 ugm.
Total 10 ugm.
17-ketosteroids 7.4 mg/510 cc.
17 ketogenic steroids 8.4 mg/510 cc.
Basal metabolism rate minus 8 (weight 134; height 63").

6-1-61: Last seen in clinic. General physical examination and pelvic were normal, however no menses had yet occurred.

Note: Stock of this case was not sufficient for the number of slides needed for this conference, therefore a histologically similar case has been substituted.

CASE NO. 15 DECEMBER 3, 1961

ACCESSION NO. 11611 OUTSIDE NO. S60-292

NAME: J. Y.
AGE: 61 SEX: Male RACE: White

CONTRIBUTOR: Donald L. Alcott
Santa Clara County Hospital
San Jose, California

TISSUE FROM: Left lung (surgery)

CLINICAL ABSTRACT:

History: This patient underwent left thoracotomy on February 17, 1960, because of an abnormal chest x-ray. A tumor was found in the left lower lobe and a left pneumonectomy was performed. No metastases were found at the time of operation.

GROSS PATHOLOGY:

In the left lower lobe bronchus was an elevated 2 x 1.3 cm. tumor which had destroyed the bronchial mucosal markings and had extended into the prehilar lymph nodes. The lung tissue distal to the tumor was engorged with blood, and the bronchi were dilated with thick yellow purulent material. Ther was a gross margin of 1 cm. between the visible tumor and the bronchial resect line, and frozen section of the surgical bronchial margin revealed no tumor. There was no gross or microscopic involvement of the separately submitted hila lymph nodes.

COURSE:

He did well until August 1960, when he began to complain of shortnes of breath. On September 24, 1960, he was admitted to the hospital because of generalized body swelling and shortness of breath.

Physical examination: There was moderate swelling of the face and arms. The neck veins were flat. There were a few soft lymph nodes in the lef supraclavicular area. There was a well healed thoracotomy scar. The left che was uniformly dull and there were no breath sounds. The liver was enlarged to 3 cm. below the right costal margin. There were healing herpetic lesions on t left leg. In the hospital the patient presented a problem in the diagnosis differential of edema. There was no history or physical evidence of renal or heart disease. Although there was swelling of the face, the absence of venous distention ruled out the possibility of superior vena caval syndrome. Venous pressure was 6 cm. measured in the right antecubital vein with the patient lying flat and arm held up to chest level. The patient's blood pressure was 90/56; pulse 102; respiration 25; and normal temperature.

ACCESSION NO. 11611

Laboratory report: On October 8, 1960, serum sodium was 153 mEq/L. serum potassium was 2.7 mEq/L. He was treated with 1000 mg. sodium diet and potassium triplex. By November 1, 1960, his serum sodium had improved to 146 mEq/L and the potassium to 3.1 mEq/L. When he was presented to Grand Rounds, the concensus of opinion was that the edema was due to excessive adrenocortical steroids. Further studies done at Stanford showed 17-hydroxysteroids averaged 135 mg/24 hr; 17-ketosteroids averaged 50 mg/24 hr. Retroperitoneal air studies revealed enlarged adrenal glands bilaterally.

A transphenoid hypophysectomy was performed on December 7, 1960, following which the patient gradually deteriorated, contracted pneumonia, and expired on December 22, 1960.

AUTOPSY:

There were metastases to the supraclavicular, hilar, and periaortic lymph nodes, both adrenals, liver, spleen, right kidney, gastrocolic ligament, and the right frontal lobe of the brain. The right adrenal weighed 105 gm. and measured 10 x 7 x 6 cm. The cortex was hemorrhagic, tumorous, and 3 cm. thick. The medulla was soft, homogeneous, firm, and yellow-gray. The left adrenal weighed 35 gm. and measured 8 x 5 x 4 cm. The section was similar to that of the right adrenal.

BIO-ASSAY:

"Lyophilized plasma was assayed for corticotropic activity in hypophysectomized rats by the method of Lipscomb and Nelson; decidedly elevated levels of 4.5 milliunits per 100 ml. of plasma were found....Assays of tumor revealed corticotropic activity of 23 milliunits per gram of tissue. The concentration of corticotropin in the pituitary was unusually low at 0.7 milliunits per mg. of tissue. Corticotropic activity was not found in other tissues. It was postulated that the carcinoma was autonomously secreting lar quantities of corticotropic substance."

Note: Thanks are extended to Drs. John Nuckton and Grant Little for the bio-assay of the tumor.

CASE NO. 16 DECEMBER 3, 1961

ACCESSION NO. 10791 OUTSIDE NO. S-1319-59

NAME: J. B.
AGE: 32 SEX: Male RACE: Caucasian

CONTRIBUTOR: Francis S. Buck
Los Angeles County Hospital
Los Angeles, California

TISSUE FROM: Left testis

CLINICAL ABSTRACT:

The patient first sought medical attention in August 1959 for a sudden enlargement of a left scrotal mass which had been present for about four months. He had had a left inguinal herniorrhaphy at the age of 22.

SURGERY:

On August 10, 1959, a left testicular tumor was removed. There was evidence then of a left inguinal lymphadenopathy.

GROSS PATHOLOGY:

The specimen was submitted in fragments, which aggregated 37 gm., and exhibited focal areas of hemorrhage and necrosis.

COURSE:

The patient entered the Los Angeles County Hospital for terminal care, on August 31, 1959. The urinary 17-ketosteroids were 23.4 mg/24 hr. (September 2, 1959); urinary 17-ketogenic steroids were 19.8 mg/2080 ml. and chorionic gonadotrophins were greater than 200,000 I.U./L (September 11, 1959). No apparent response in the hormonal levels were noted following an attempt at Methotrexate therapy. The patient expired on September 13, 1959.

Autopsy revealed very extensive pulmonary metastases and a solitary metastatic nodule in the liver.

CASE NO. 17 DECEMBER 3, 1961

ACCESSION NO. 10914 OUTSIDE NO. HPS-5953-59

NAME: L. D.
AGE: 25 SEX: Female RACE: White

CONTRIBUTOR: S. K. Abul-Haj
Walter Reed Hospital
Washington, D. C.

TISSUE FROM: Left adnexal mass

CLINICAL ABSTRACT:

History: This married, 25 year old female sought medical attention because of irregularly irregular menses, infertility, and excessive vaginal bleeding. These symptoms had occurred over a period of 1½ years, characterized by periods of amenorrhea of 2-3 months' duration and menometrorrhagia without regular pattern. The menses, when they occurred, lasted 10-12 days and the intermenstrual intervals varied from 7-14 days. At times the vaginal bleeding was excessive and at other times it was spotty and intermittent. No other symptoms were present.

Physical examination was unremarkable. Pelvic examination revealed a serosanguinous material exuding from the cervical os. A large, ovoid, firm, discrete, left adnexal mass was felt.

COURSE:

The pregnancy test was negative. Dilatation and curettage showed hyperplastic, proliferative, secretory, and desciduoid endometrium thought to be consistent with the pattern of hormonal irregularity.

SURGERY:

In June 1959, an exploratory celiotomy was performed and a large ovoid, encapsulated left adnexal mass was removed.

GROSS PATHOLOGY:

The specimen consisted of an ovoid, firm, rubbery, well encapsulated mass, measuring 8 x 10 cm. The surface was smooth and shiny. The cut surface revealed it to be composed of interlacing trabeculae of gray-white fibrous tissue interspersed with irregular islands of homogeneously finely granular soft yellow tissue. These yellow areas were oily to touch. A thin rim of ovarian stroma was also present, which contained several small follicular retention cysts and an edematous myxoid stroma.

ACCESSION NO. 10914

FOLLOW-UP:

When the patient was seen in November 1959, her menses were regular but on a 21 day cycle rather than the previous normal of 28 days. No complaints or significant findings were present.

CASE NO. 18 DECEMBER 3, 1961

ACCESSION NO. 11413 OUTSIDE NO. Unknown

NAME: Unknown
AGE: 70 SEX: Male RACE: White

CONTRIBUTOR: Karl E. Kirschner
San Luis Obispo, California

TISSUE FROM: Lower Lobe of right lung

CLINICAL ABSTRACT:

In January 1961 the patient was seen by his physician because of several months of increasing tiredness, difficulty in awakening in the morning, and dizziness before meals. Except for a posterior myocardial infarction seven years previously, past medical history was unremarkable. On the morning of January 8th his wife was unable to arouse him and he was admitted to the hospital in semistupor. On admission it appeared as though he was in congestive failure and a chest film revealed marked cardiac enlargement and pleural fluid bilaterally. Oxygen was given and he responded rapidly leaving the hospital the next day. A few days later he was again admitted, this time in deep stupor with salivation, profuse perspiration, a flushed face and stertorous breathing. The blood sugar examination revealed only 54 mg% reducing substance. The patient responded dramatically to intravenous glucose. Further laboratory studies included total 17 ketosteroids of 7.4 mg/24 hr and 11.3 mg/24 hr of 17 hydroxycorticosteroids. Following ACTH stimulation the 17 ketosteroids rose to 22 mg/24 hr and the 17 hydroxycorticosteroids rose to 25.5 mg/24 hr. No abdominal masses were palpated and the general physical and routine laboratory studies were not remarkable. A repeat chest film showed a space occupying lesion in the right lower chest.

SURGERY:

A thoracotomy was performed on January 20, 1961. The entire right lower lobe was replaced by a firm elastic tumor which apparently had not invaded the lung but appeared to have compressed the lobe into a thin shell of atelectatic parenchyma at its base. A lobectomy was performed.

GROSS PATHOLOGY:

The specimen weighed 1250 gm. and measured 25 x 15 x 18 cm. An anthracotic pigmentation was noted over almost the entire smooth surface. On section, the neoplasm was trabeculated, light tan, focally cystic, and not unlike that of a large degenerated uterine fibroid.

COURSE:

Following surgery, the blood sugar levels were 150 mg%, the next day 370 mg%. Following insulin medication the sugar level dropped to 150 mg%, 145 mg%, 95 mg%, and 150 mg% during the consecutive days. The patient was discharged in good condition and is now free of symptoms presenting blood values within the normal range for the past month. Repeat chest x-ray disclosed no recurrent pulmonary infiltrates. The patient has not received insulin since January, 1961.

CASE NO. 19 DECEMBER 3, 1961

ACCESSION NO. 11489 OUTSIDE NO. S-61-1299

NAME: B. T.
AGE: 35 SEX: Female RACE: Caucasian

CONTRIBUTOR: S. K. Abul-Haj
Walter Reed Hospital
Washington, D. C.

TISSUE FROM: Right ovary

CLINICAL ABSTRACT:

History: This patient sought medical help approximately one year ago for amenorrhea of two years' duration. There were no other complaints. At that time the work-up was unrevealing. It included an endometrial biopsy which was reported as insufficient for diagnosis. The remainder of the history and family history were noncontributory. She was referred to this hospital for definitive diagnosis.

Physical examination: The examination was essentially within normal limits except for a right adnexal mass.

Laboratory report: Routine hemograms and urinalysis were within normal limits.

SURGERY:

On March 8, 1961, a celiotomy with right salpingo-oophorectomy was performed. A rounded right ovarian mass, 5.5 cm. in diameter,was found. It was easily removed together with the right salpinx to which it was firmly attached. She had an uneventful post-operative recovery.

GROSS DESCRIPTION:

The specimen consisted of an ovoid ovarian mass, measuring 6.0 x 6.0 x 5.0 cm., with the fallopian tube attached. The surface was smooth, pearly-white, except where the fallopian tube was plastered to it by fibrous stringy adhesions. The cut surface was finely granular, yellow-tan to orange-yellow with small irregular areas of cystic hemorrhagic liquefaction. The mass was completely surrounded by an intact ovarian capsule and an ovarian gray-white cortical stroma which varied in thickness from area to area. There were small nodular growths within the ovarian stroma and separated from the main mass by a thin rim of enveloping cortical stroma. The nodules were, however, completely contained within the ovary. Multiple sections showed no extension outside

the ovary.

FOLLOW-UP:

To date (10-2-61) patient is free of disease. She resumed menstruation six weeks after removal of the ovarian mass, and her periods have been regular.

CASE NO. 20 DECEMBER 3, 1961

ACCESSION NO. 9849 OUTSIDE NO. S-56168
LACGH NO. 59723

NAME: M. T.
AGE: 51 SEX: Female RACE: Caucasian

CONTRIBUTOR: J. E. Kahler
St. Vincent's Hospital
Los Angeles, California

TISSUE FROM: Right adrenal tumor (surgery)

CLINICAL ABSTRACT:

History: The patient was born in Yugoslavia and emigrated to the U.S. A. in early 1957. Menarche occurred at the age of 12 and she had normal regular menses until 1945 (age 39) when she developed amenorrhea while in a concentration camp. Normal menses were reinstituted after a three year interval and continued normally for six more years until 1954 (age 48) when she experienced complete amenorrhea without oligomenorrhea or other menopausal symptoms.

For the past two years the patient had noted growth of hair on the chin, chest, and abdomen; for the last eight months she had experienced shortness of breath, ankle edema, and hypertension.

Physical examination: When the patient was admitted to the hospital on October 30, 1957, the following positive findings were recorded: Blood pressure 210/130; pulse 80, regular. The heart and the liver were enlarged. There was a grade IV pitting edema of both ankles. No adrenal masses were palpated. Hirsutism of the face, trunk, and extremities were noted, but she did not have striae.

X-ray report: Retroperitoneal air injection studies demonstrated a tumor, measuring 6 cm. in diameter, in the right adrenal area. A solitary metastasis to the second rib was confirmed subsequently by biopsy.

Laboratory report: Regitine test negative (maximum reduction 8-13); Thorne test negative (reduced from 62 to 5/cu.mm); Noradrenaline 3 ug/100 ml. (normal up to 14); 17 Ketosteroids 32 mgm/24 hr output urine (normal 6-15); 11 Oxysteroids 28 mgm/24 hr output urine (normal 5-18); Urine potassium 1.35 gm/Liter (normal 1-3 gm/day); Urine sodium 0.88 gm/Liter (normal 3.5 gm/day); Serum potassium 16 mgm (normal 16-22 mgm); Serum sodium 345 mgm (normal 300-345 mgm); Glucose tolerance: Fasting 92, ½ hr 140, 1 hr 160, 2 hr 235, 3 hr 170. All urine sugars free; Hb 16 gm., Rbc 4.8 million, Wbc 7,100, normal differential; Urine 4+ albumen, occasional hyaline cast. 15-20 Wbc, 2-4 Rbc.; NPN 45 mgm.

SURGERY:

On November 21, 1957, a right adrenalectomy was performed. The large tumor extended to the opposite side, passing under the aorta and the inferior vena cava. The tumor was incompletely removed. The metastases in the liver were already evident.

GROSS PATHOLOGY:

The ovoid right adrenal mass measured 7 x 6 x 5 cm. and weighed 95 grams. The capsule was not intact and the tumor had broken through it and extended into the adjacent fat. On section, the tumor was soft, orange-yellow, mottled with white infarcts and red areas of hemorrhage. No normal adrenal tissue could be grossly identified.

COURSE:

The patient entered the Los Angeles County Hospital for palliative treatment. She had a large, firm, nontender, purple bleeding mass on the right side of the mouth which displaced the tongue to the left. A firm, nontender, fixed mass, measuring 7 cm., was present on the anterior chest above the right breast, and another firm mass, measuring 6 cm., was found in the posterior left thorax near the waist. The x-rays of the chest and mandible revealed metastatic lesions. On February 24, 1958, urinary 17-ketosteroids were 69.2 mg/900 ml. and 17-ketogenic steroids (Norymberski) were 64.5 mg/900 ml. (total 24 hr. urine volume was 900 ml.). Pregnanediol 6.9 mg/24 hr. and pregnanetriol 11.5 mg/24 hr. (normal up to 3 mg.).

The patient died on March 19, 1958. Autopsy (LACH 59723) showed no remnants of the right adrenal except some scar tissue, an atrophic left adrenal (weight 2 gm.) and extensive metastases to the liver, lung, bone, including mandible.

CASE NO. 21 DECEMBER 3, 1961

ACCESSION NO. 11713 OUTSIDE NO. S-1895-59

NAME: M. V.
AGE: 20 SEX: Female RACE: Caucasian

CONTRIBUTOR: Francis Buck
Los Angeles County Hospital
Los Angeles, California

TISSUE FROM: Uterus

CLINICAL ABSTRACT:

This gravida III, para I, 20 year old female delivered a normal full term male infant on September 17, 1959. Labor and gestational course were not remarkable. The placenta was not remarkable. One month later, vaginal bleeding necessitated therapeutic uterine curettage. Products of conception were reported seen in the specimen. Two weeks later, persistent vaginal bleeding again necessitated hospitalization and treatment. A positive pregnancy test was obtained a week later after the patient had been discharged. On November 24, 1959 she was admitted to the Los Angeles County Hospital in shock with profuse vaginal bleeding. Patient was stablized and transfused. Physical examination revealed abdominal pain. Uterine curettage and culdoscopy were performed. On the next day profound shock again developed and an emergency laparotomy was performed. A large ruptured cyst of the right ovary and an intra-abdominal hemorrhage were found. The left ovary also appeared cystic and the uterus was enlarged to the size of a two months' gestation. A right salpingo-oophorectomy and a left ovarian cystectomy were performed. On this day the report of the dilatation and curettage was given to the physicians. Laboratory studies were ordered.

Laboratory and X-ray reports: Urinary chorionic gonadotrophin titer, using animal assay, was found greater than 10,000 but less than 40,000 international units per liter. The serum leucine amino peptidase level was 172 (female normals are 80-210). A post-laparotomy chest x-ray revealed bilateral pulmonary infiltrations.

SURGERY:

On December 12, 1959, a hysterectomy and a left salpingo-oophorectomy were performed.

GROSS PATHOLOGY:

Hemorrhagic, friable necrotic tumor was found in the uterus and fallopian tube.

ACCESSION NO. 11713

COURSE:

Postoperative (2½ weeks) gonadotrophin levels were reported as being between 40,000 and 200,000 I.U. Methotrexate therapy was begun and levels were less than 10,000 on January 4, 1960. Within 5 weeks after institution of therapy, the assays were negative until September of 1961. At that time the assays were reported as over 1,000 but under 10,000 I. U. per liter. Chest x-ray on June 13, 1961 was negative.

Chorionic Gonadotrophin Titers. Performed by the Hyperemic Ovarian Response Animal Assay Method under the direction of Benjamin Horwitt, Ph.D. at Bio-Science Laboratories in Los Angeles.

CASE NO. 22 DECEMBER 3, 1961

ACCESSION NO. 11820 OUTSIDE NO. A-121-48

NAME: M. K.
AGE: 11 mo. SEX: Female RACE: Caucasian

CONTRIBUTOR: Eva Heuser
Childrens Hospital
Los Angeles, California

TISSUE FROM: Adrenal

CLINICAL ABSTRACT:

History: This child was born on January 7, 1948 at full term by podalic version of a breech presentation. Her birth weight was 7 pounds 6 ounces and at seven days, 7 pounds 15 ounces. There were no neonatal complications. The weight gain was unsatisfactory on breast feeding, so full similac formula was started. Weight gain then was satisfactory (14 ounces in one week). At four weeks vomiting of some feedings occurred. It was not projectile and contained no blood or bile. There was no diarrhea. At five weeks of age, the child was admitted to Childrens Hospital for a complete workup because of feeding problems and infections. On April 22, 1948, because of continued vomiting, fever in spite of adequate antibiotics, and electrolyte disturbances Desoxycorticosterone acetate (0.5 mgs. daily) was started. Rapid weight gain ensued with generalized improvement in clinical appearance. On May 17, 1948 steroids were decreased to 0.25 mg. daily and the patient was discharged on 7 gms. added salt to the diet. She did well until October 29, 1948 when vomiting again developed and food was refused. On November 12, 1948 she was admitted to the hospital in an irritable state with cold extremities.

Laboratory report: On her first admission the hemoglobin varied from 14.7 to 9.0 gms. and two transfusions were given. The white blood count varied from 7,850 to 14,950. The urine showed a trace of albumin and an occult to 25 pus cells consistently until May 12, 1948. After that time urines were negative. Serum sodium on February 18 was 330 mg.% and on April 10 was 305 mg.% (normals 320-350 mg.%); serum potassium on Feb. 18 was 29 mg.% (normal 15-20 mg.%); serum chlorides on April 10 was 522 mg.% (normal 570-620 mg.%); non-protein nitrogen on Feb. 24 was 40 mg.%; blood sugar on Feb. 24 was 99 mg.%; and 17 ketosteroids on Feb. 26 were 6.1 mg/100 ml. or 5.1 mg/24 hr (normal at this age 1.0 mg/24 hr.)

X-ray reports: Cystogram revealed no urogenital sinus or fistula. On March 5, 1948 a chest x-ray revealed density in both upper lung fields consistent with bronchopneumonia. On March 16, the chest had cleared. IVP was unsatisfactory and retrograde pyelogram revealed no abnormalities. On the same day no carpal bones were observed. By May 7 one carpal bone was noted, and on June 9, two were present.

An EKG showed marked right axis deviation and sinus tachycardia.

COURSE:

On the day following admission the temperature spiked to 102.6. Drowsiness, irritability and vomiting increased, shock developed in spite of therapy and the child died. Terminally black material was vomited.

AUTOPSY:

The body was that of an 11 month old, well developed, and well nourished, white female measuring 78 cm. in length (normal length 70 cm.). The skin showed a peculiar yellow-brown discoloration about the external genitalia, anus, and axilla. There was a moderate amount of light brown pubic hair present over the mons pubis and the labia majora. The clitoris was noted to be markedly enlarged and measured 1.3 cm. in diameter and 2.8 cm. in length. The breast and nipples were not remarkable. The features were noted to be adult in configuration, the cheeks having lost the fatty pads of infancy and the nose having become more prominent. The extremities appeared elongated in relation to the torso.

The organs in both cavities were in their normal arrangement and appeared essentially normal. There was no fluid in either cavity.

Adrenals: Both of the adrenals were markedly enlarged. The left adrenal weighed 14.6 gms. and measured 5.8 x 4.0 x 3.8 and 1.2 cm. in greatest dimensions (normal 4 to 5 grams each). The right adrenal weighed 11.7 gms. and measured 4.0 x 5.4 x 3.0 and 1.4 cm. in greatest dimensions. They occupied their usual anatomic positions at the superior poles of the kidneys completely capping the superior one-half of each kidney. On palpation the organs were firm, rugose in contour, and no nodules could be felt. The cut surfaces revealed a pale yellow outer cortical layer and a darker brown inner medullary layer. These layers appeared to be in approximately normal proportions for an adult adrenal. The preponderance of pale yellow cortical tissue usual at this age could not be made out.

Ovaries: The left ovary measured 1.3 x 0.3 x 0.2 cm. The right ovary measured 1.2 x 0.4 x 0.2 cm. In the right ovary a small translucent cystic appearing structure was noted. In the perirenal fat and along the course of the ureters in the retroperitoneal tissue, several small discrete nodules of pale yellow brown tissue were noted.

External Genitalia: A typical picture of pseudohermaphroditism was presented. The labia majora were markedly enlarged and appeared scrotal in character. Upon dissecting along the inguinal canal, a round ligament was found which broke up into numerous fine fibrils which were spread diffusely through the labia majora. No labia minora could be made out, although there was a loose tissue which resembled a prepuce around the clitoris and extended for a distance of approximately one-half cm. along the lateral borders. The urethra opened behind the clitoris along the anterior vaginal wall. The vagina was hypoplastic, but was approximately normal in contour.

CASE NO. 23 DECEMBER 3, 1961

ACCESSION NO. 11847 OUTSIDE NO. S61-1112

NAME: K. H.
AGE: 43 SEX: Male RACE: Caucasian

CONTRIBUTOR: Gerson R. Biskind
Mt. Zion Hospital
San Francisco, California

TISSUE FROM: Right adrenal

CLINICAL ABSTRACT:

Patient was hospitalized in February 1961, following an auto accident. Examination showed centripetal obesity, prominent purplish striae over abdomen, buffalo hump. His chief complaint was pain in the back, muscular weakness and loss of libido leading to divorce. His entry blood pressure was 150/100, and electrocardiogram suggested left ventricular hypertrophy. The visual fields and fundi were normal. STS positive. X-rays: Sella and IVP normal. Routine laboratory examinations and liver function studies were normal. Pre-operative steroid studies were as follows:

	17 KS	KGS	PS	
Feb. 1961	7.3	31.3	14.1	Control
Mar. 1961	11.4	25.2	28.2	Suppression with dexamethosone
Mar. 1961	14.5	61.6	29.6	After ACTH
Apr. 1961	5.5	26.6	11.7	After SU 4885

SURGERY:

On April 31, 1961, an abdominal exploration was done, and a distinct mass was palpated in the right adrenal; the left adrenal was interpreted as small or atrophic on palpation.

GROSS PATHOLOGY:

The entire right adrenal was removed. It contained a circumscribed encapsulated nodule, 2 cm. in diameter. The remaining adrenal was atrophic.

COURSE:

He was discharged on 30 mg. of cortisol daily; no ACTH was given. The amount of cortisol was progressively decreased without discomfort until he was supported on 5 to 10 mg. daily. The patient left the area and was lost to follow-up until August 1961 when he re-entered the hospital complaining of severe weakness of the leg muscles, requiring crutches for locomotion.

ACCESSION NO. 11847

He had been unable to resume work because of weakness. The amount of cortisol taken varied greatly; the patient increased the amount in an attempt to overcome the weakness, but without success. The present examination included steroid studies listed below after he had not taken cortisol for 3 to 7 weeks. The following x-rays were negative: Sella, chest, retroperitoneal air insufflation, and I.V.P. The striae and buffalo hump were unchanged.

Treatment at present is limited to testosterone for possible improvement in muscular strength.

Post
~~Pre~~-operative steroid studies:

	17 KS	KGS	PS	
Aug. 23, 1961	6.1	27.5	-	Control
Aug. 30, 1961	6.3	27.0	11.7	Control
Sept. 3, 1961	7.7	18.5	13.4	After dexamethosone, 8 mg/day, 2 days
Sept. 5, 1961	8.1	24.4	18.3	Control
Sept. 6, 1961	11.2	42.0	23.5	p̄ ACTH
Sept. 16, 1961	6.3	22.5	18.6	Control
Sept. 17, 1961	7.5	20.3	17.2	After SU 4885
Oct. 3, 1961	9.4	20.2	13.7	Control
Oct. 11, 1961	7.6	18.0	13.2	Control
Oct. 16, 1961	4.5	16.4	13.5	Control

CASE NO. 24 DECEMBER 3, 1961

ACCESSION NO. 11857 OUTSIDE NO. 61-12056 *

NAME: G. L. K.
AGE: 34 SEX: Female RACE: Caucasian

CONTRIBUTOR: Ernest Courier
Glendale Memorial Hospital
Glendale, California

TISSUE FROM: Left adrenal

CLINICAL ABSTRACT:

History: This patient was admitted to the hospital on January 3, 1961, with a history of a weight gain of 50 lbs. over the past two years, together with weakness, left flank pain, and double vision of eight months' duration. She stated she was forced to quit her work as a registered nurse eight months ago because of the increase in weakness. For the past eight months, the weight gain had been limited to the face and abdomen with the onset of a moon facies, but prior to that the weight gain had exhibited a generalized distribution. The left flank pain was dull, aching, and somewhat worse on standing. The past history included rheumatic heart disease at age 12, intestinal obstruction at age 15, which was surgically treated, and removal of a large ovarian cyst with simultaneous hysterectomy at age 22 years.

Laboratory examination: Hemoglobin was 11.6 grams with hematocrit 34. Serum electrolytes were within normal limits, save for a serum calcium which was repeatedly found to be in the range of 8.5 mg.%. 17 ketosteroids 53.2 mg./24 hrs., and 17 ketogenic steroids 33.1 mg./24 hrs. Dehydroepiandrosterone levels were 1.4 mg./24 hrs. Urine creatinine 2.4 grams/24 hrs.

X-ray films of the spine and pelvis were negative and an intravenous pyelogram was entirely normal with no evidence of adrenal enlargement.

SURGERY:

On February 4, 1961, a left lumbar adrenalectomy was performed. There was no evidence of tumor or hyperplasia of the gland.

GROSS PATHOLOGY:

The gland weighed 6.5 grams and measured 5.5 x 2.5 x 1.5 cm. when stripped of fat. The cortex was 2 mm. in thickness throughout the

gland and the medulla was grayish in color and non-hemorrhagic.

Microscopic examination revealed a histologically unremarkable adrenal gland.

COURSE:

Postoperatively, the patient's electrolytes remained satisfactory and her condition gradually improved. 17 ketosteroids determined postoperatively, were 8.0 mg./24 hrs., and 17 ketogenicsteroids 19.4 mg. per 24 hours.

*Normal gland removed from patient at time of surgery was inadvertantly discarded, and sufficient material was not left for the conference. Therefore surgically removed normal adrenal from files of a Los Angeles County Hospital case was used.

CASE NO. 25 DECEMBER 3, 1961

ACCESSION NO. 11901 OUTSIDE NO. 61-2189

NAME: T. W. K.
AGE: 49 SEX: Male RACE: Caucasian

CONTRIBUTOR: G. R. Biskind
Mt. Zion Hospital
San Francisco, California

TISSUE FROM: Left testis

CLINICAL ABSTRACT:

History: The patient entered the hospital with a complaint of swelling and tenderness of the left testicle which had persisted for three months following trauma. The nature and severity of the trauma was not specified. Past history revealed previous bouts of amoebic dysentary and malaria but was otherwise unremarkable.

Physical examination: Blood pressure was 140/70 and the general physical was unremarkable except for the left testis which was twice the normal size and hard. The right testis appeared normal.

Laboratory report: Gonadotrophins on June 20, 1961 were positive at 50 units, but negative at 100, 200, 500, and 1000 units. Chest x-ray was negative. I.V.P. showed lateral deviation of the left ureter 4 - 5 cm. below the ureteropelvic junction.

SURGERY:

On June 22, 1961, a left orchiectomy was done, followed a week later by a retroperitoneal node dissection.

GROSS PATHOLOGY:

The testis weighed 70 gm. and there were large areas of yellowish necrosis alternating with friable and hemorrhagic gray tumor tissue. Nodes submitted were large and necrotic in appearance.

COURSE:

Following surgery, the patient received three 5 day courses of Actinomycin D plus methotrexate and leukeran. Gonadotrophin studies done on July 21, 1961 were positive at 5 units, but negative at 10, 30, and 60 units Repeat studies on September 6, 1961 were positive at 5 units, but negative at 50 and 100 units.

FOLLOW-UP:

As of November 2, 1961, the patient was alive with no evidence of recurrence, and chest x-ray and I.V.P. films were normal.

USEFUL LABORATORY TESTS FOR EVALUATION OF ADRENAL STATUS

CLINICAL CONDITIONS: 1. Normal; 2. Cortical Atrophy or Necrosis; 3. Panhypopituitarism; 4. Hyperthyroidism; 5. Hypothyroidism; 6. Gout; 7. Hirsutism

State of Adrenals	Basal Level PS*	Basal Level KGS**	Basal Level 17 KS	ACTH Stimulation PS	ACTH Stimulation KGS	ACTH Stimulation 17 KS	SU 4385 30 mg./kg./4hr. IV	Suppression Dexamethasone a) 0.5mg. 4id b) 2.0mg. 4id	Plasma F-S Levels
Normal Adult	0.05 mg./ kg. body wt.	0.1 mg./ kg. body wt.		3-5X increase			2X increase PS. - no change	50% of normal output	10-20 mcg.% at 8 AM 5-10 mcg.% at 5 PM
Primary Cortial Atrophy or Necrosis	Very low (5% may be normal)			D- SI	D- SI	D- SI	No response		
Secondary Atrophy (Cortisone)	Very low			Pt. maintained on same dose of cortisone, given ACTH ger 5 days, compare 1st & 5th day (Addison's: no rise)			No response		
Panhypopituitarism	Very low			2-3X increase			No response		
Hyperthyroidism	May be 2X normal			3-5X increase			2X increase		normal 10-20 mcg.%
Hypothyroidism	Low						2X increase		
Gout	Low			3-5X increase			2X increase		
Hirsutism	Normal	Normal	Upper limit normal	3-5X increase					
	Normal	Slight incr. ***	Upper limit normal	3-5X increase					

Porter Silber Chromogen

*PS: 17-21 dihydroxy 20 ketosteroids or 17 hydroxy corticosteroid

**KGS: Ketogenic steroids

***: If F increased, congenital adrenal 3 hyperplasia possible.

Blue pretibial skin pigmentation

CUSHING'S SYNDROME

Pure Metabolic; Mixed pattern with virilization or demasculinization:

Adrenal Lesion	Basal Level			ACTH Stimulation			SU 4885			Suppression Dexamethasone			X-ray with or without Gas Insufflation
	PS	KGS	17 KS	PS	KGS	17 KS	PS	KGS	17 KS	PS	KGS	17 KS	
None	I	I	I	I	I	I	D	I	N	D	D	D	No masses
Hyper-plasia	I	I	N-SI	I greater than 5X	I	I	D-NC	I	I	NC* D**	NC* D**	NC* D**	Bilateral masses
Adenoma	I	I	D-N	I	I	I	NC	I-NC	I-NC	NC* NC**	NC* NC**	NC* NC**	Unilateral mass
Carcin-oma	I	I	I	NC	NC	NC	NC	NC	NC	NC**	NC**	NC**	Unilateral mass
Extra-adrenal tumors	I	I	I	I-NC	I-NC	I-NC	NC	NC	NC	NC	NC	NC	

N = Normal
D = Decreased
I = Increased
S = Slightly
NC = No change

* 2 mg./day for 3 days
** 8 mg./day for 2 days

ADRENO-GENITAL SYNDROME

Including: Virilization in Female, congenital or acquired, with or without pseudohermaphroditism; ?
Virilization in Male; Feminization of Male; Simple Hirsutism; ?Stein-Leventhal Syndrome

Adrenal Lesion	Basal Level				ACTH Stimulation				SU 4885				Suppression with 9 FF				X-ray with or without Gas Insufflation
	PS	KGS	17 KS	P_3	PS	KGS	17 KS	P_3	PS	KGS	17 KS	P_3	PS	KGS	17 KS	P_3	
Hyperplasia	SD or N	I	I	Cg. I Ac. N	SI	SI	SI	SI	D	I	I	SI	D	D	D	D	Bilateral masses
Adenoma	D-N	I	I	N-I	NC-SI	NC-SI	NC-SI	NC-SI	D	NC	NC	NC	NC	NC	NC	NC	Unilateral mass
Carcinoma	D-N	I	I	I	NC	NC	NC	NC	NC	NC	NC	NC	NC	NC	NC	NC	Unilateral mass
Virilizing Tumors, not Adrenal**	N	N-I	N-I	N	NC*	NC*	I*	NC*	NC*	NC*	NC*	NC*	NC*	NC*	NC*	NC*	

N = Normal
D = Decreased
I = Increased
S = Slightly
NC = No change

P_3 = Pregnantriol
PS = Porter-Silber Chromogens
KGS = Ketogenic steroids
17 KS = Neutral 17 ketosteroid

* Response of normal adrenal may mask
** Severe virilization by testosterone from tumor not reflected in 17 KS or KGS assays

Cg.= Congenital
Ac.= Acquired