[Combined Case reports

polycystic Kidney diseases, hypertensive encephalopathy, chronic obstructive pulmonary disease And obesity]

GRAND ROUNDS

Case for [redacted] 1958 - Dr. Edith Potter has consented to discuss

[redacted] was a white female infant that was admitted to [redacted] at the age of 23 month on [redacted] 1954. Four months previously the patient had developed scarlet fever, following which there was a history of failure to thrive and to gain weight satisfactorily. One month prior to admission it had been thought by medical opinion that the patient was anemic and two blood transfusions were given. Subsequently and over the following weeks the patient became lethargic, and "would only sit." On the day prior to admission the child had a generalized seizure characterized by incontenance and generalized twitching which lasted for one minute and was followed by vomiting. The pediatrician admitted the patient to the hospital in [redacted] Texas, where a spinal puncture was done under G.A. Pressure = 360 mm H_2O and cell count was 10 lymphocytes. The patient was then transferred to [redacted] with the tentative diagnosis of brain tumor.

Physical Examination: Normal temperature and pulse but respirations 50 per minute. Weight only 16 pounds. Blood pressure was not taken. Patient was described as both acutely and chronically ill with wasting. A mottled appearance was present in the skin and echymosis 2 cm. x 2 cm. was present over the right eyebrow. Tissue turgor was diminished; tongue dry.

Eyes: Pupils equal - reacted to light

Fundi: Bilateral papilledema (no hemorrhages)

Chest: Respirations were rapid with retraction of the lower ribs. (Presumably the patient could have been described as having air hunger or acidotic breathing.

Heart: Nothing abnormal demonstrated

Pulse pressure was satisfactory by feel.

Abdomen: Liver edge 2 fingers below costal margin. No masses felt. Rectal examination revealed no pelvic mass.

Neurological Examination: Revealed a 6th nerve palsy to the left and some trismus of the masseter muscle. Some suggestion of carpo-pedal spasm. Deep tendon reflexes were absent.

Laboratory Studies:

RBC 4 million, Hb - 11.8, WBC - 35,500, differential WBC - 53 polys, 3 immature polys, 44 lymphocytes.

Urine: Albumin trace, 1 red cell per high powered field, few epithelial casts.

CO_2 - 8.2 mEq.

Cl - 105 mEq/L

Blood culture - negative

Throat culture - Staph aureus

Course in Hospital: Patient placed in oxygen. It was anticipated that the tumor was present in the posterior fossa. I.V. fluids given with Ca gluconate. Acidotic breathing relentlessly continued. Skull and chest x-ray showed no abnormality. Five and one half hours after admission cardiac and respiratory arrest occurred.

Autopsy: Kidneys weighed 20 grams each, measured 6 x 4 x 2½ cms. Grossly polycystic kidneys - renal architecture completely destroyed by cysts of various sizes (3/4 cm. to microscopic). The findings suggested a combined type of disease, i.e. - cysts resulting from embryonic failure of the connecting tubules to join with the proximal portions of the nephrons and a second type of cyst due to dilatation of the collecting tubules and intra-aterine obstruction of the ureters.

The pericardial sack was grossly distended with fluid (50 cc of gray-brown fluid).

Lungs: right lung was mottled grey and brown with diminished crepitance. The cut surface showed brownish pink color and bleeding was scanty.

Brain: Normal

Histology: Kidney - two types of cysts (1) Dilated Bowman's Capsule, containing glomerular buds, composed of capillary endothelial cells. (2) Cysts from tubular areas - lined by tubular epithelium. Cysts free of content.

Lungs - revealed areas of focal aletectasis. Some small focal areas of pulmonary edema.

Diagnosis - Polycystic kidneys with hypertensive encephalopathy

MEDICAL GRAND ROUNDS

MAY 14, 1958

CASE 1. [redacted], A 35-YEAR-OLD WHITE MALE [redacted], WAS FIRST ADMITTED TO [redacted] ON [redacted]56 BECAUSE OF ACUTE BULBO-SPINAL POLIOMYELITIS MANIFESTED BY DIFFICULTY IN HANDLING ORAL SECRETIONS, WEAK VOICE, AND WEAK COUGH. ON ADMISSION T. 102°, R 22, CO2 25, CSF 117 WBC (70% LYMPHOCYTES). VC OF 3.7 LITERS FELL TO 1.8 BY SECOND HOSPITAL DAY. HIS LONG HOSPITAL COURSE WAS COMPLICATED BY ASPIRATION PNEUMONIA AND RECURRENT EPISODES OF SUDDEN CYANOSIS AND HYPOTENSION WHICH WERE RELIEVED BY IPPB. OTHER THERAPY INCLUDED TRACHEOSTOMY, CONSTANT NEBULIZATION OF WETTING AGENTS, BRONCHODILATORS PER IPPB, COF-FLATOR, ROCKING BED, AND ANTIBIOTICS, PRIOR TO DISCHARGE [redacted]/56, WHEN THE VC WAS 2.3 AND THE 0.5 SEC. EC 1.5 LITERS.

IN [redacted] 1957, HE DEVELOPED SOMNOLENCE, EARLY A.M. CONFUSION, PUFFINESS OF EYES, AND PERIODIC CYANOSIS DURING SLEEP. PULMONARY FUNCTION STUDIES AT THIS TIME REVEALED HYPOXIA, CO2 RETENTION, AND HYPOVENTILATION WHEN QUIET AND RELAXED, BUT NORMAL OXYGEN SATURATION WHEN ACTIVE, THOUGH pCO_2 WAS STILL ELEVATED. DETAILED STUDIES ARE ATTACHED. HE WAS THEN STARTED ON DEXEDRINE SPANSULES, 15 MGM. B.I.D., AND IMPROVED TO THE POINT THAT THERE WAS NO CHRONIC CO2 RETENTION (SECOND STUDIES).

IN [redacted], 1957, PNEUMONIA DEVELOPED REQUIRING REHOSPITALIZATION. FOLLOWING THIS DEXEDRINE WAS CONTINUED, 15 MGM. SPANSULES DAILY, AND HE SLEPT IN A SACK RESPIRATOR AT NIGHT. ON RE-EVALUATION IN [redacted] 1958, VITALOMETRY REVEALED VC 3.4 LITERS AND 0.5 SECOND EC 2.3 LITERS, BUT HYPOXIA AND CO2 RETENTION WERE AGAIN NOTED TO DEVELOP DURING SLEEP.

CASE 2. [redacted] A 49-YEAR-OLD WHITE MALE [redacted], HAD A LONG AND COMPLICATED HISTORY OF PULMONARY TUBERCULOSIS DATING BACK 26 YEARS. THIS INCLUDED A RIGHT THORACOPLASTY, CHRONIC EMPYEMA ON THE RIGHT WITH OPEN TUBE DRAINAGE, MARKED OBLITERATIVE FIBROSIS -- ESPECIALLY ON THE RIGHT, A LONG COURSE OF CHEMOTHERAPY, A RIGHT PNEUMONECTOMY 8 YEARS PREVIOUSLY, RECURRENT DYSPNEA AND PULMONARY INFECTIONS, SYSTEMIC HYPERTENSION, AND, MORE RECENTLY, COR PULMONALE TREATED WITH DIGITALIS AND DIURETICS. IN ADDITION IN RECENT MONTHS HE HAD DEVELOPED SOMNOLENCE, INCREASING IRRITABILITY, DYSPNEA, AND WEAKNESS, TWITCHING, AND MENTAL CONFUSION. HE WAS ADMITTED TO [redacted] ON [redacted]-58 FOR STUDY. IN ADDITION TO OLD PNEUMONECTOMY SCARS AND DEFORMITY WITH DRAINAGE TUBE ON THE RIGHT, THERE WERE SCATTERED RHONCHI AND TUBULAR BREATH SOUNDS OVER THE LEFT CHEST. BP 140/90 TO 200/130, P. 80, R 16 AND IRREGULAR. HEMOGLOBIN 16.5, HEMATOCRIT 65%, POTASSIUM 3.1 TO 4.9, CO2 37 TO 40. ECG SHOWED P PULMONALE AND RIGHT VENTRICULAR STRAIN. FOLLOWING PULMONARY FUNCTION STUDIES, HE WAS DISCHARGED ON SERPASIL, RITALIN, DIGITOXIN, NEBULIZED BRONCHODILATOR PER IPPB Q.I.D., AND TO SLEEP IN A SACK RESPIRATOR NIGHTLY AT 25 MM. PRESSURE. HE CONTINUES TO EXHIBIT VERY INTRIGUING TRANSIENT EFFECTS OF IMMEDIATE CHANGES IN pCO_2, BECOMING SOMNOLENT PROMPTLY WITH EITHER A HYPER- OR HYPOVENTILATION.

CASE 3. [redacted] A 42-YEAR-OLD WHITE MALE WAS FIRST ADMITTED TO [redacted] IN 1946 AT AGE 30. HE HAD "ALWAYS" BEEN OVERWEIGHT AND HAD HAD "BLUE LIPS" SINCE AGE 20, BUT HAD GAINED WEIGHT RAPIDLY SINCE DIVORSE AND "UNDESIRABLE" ARMY DISCHARGE IN 1944. HE COMPLAINED OF LEFT SUBCOSTAL PAIN, WHEEZING, DYSPNEA, AND MILD PEDAL EDEMA. WEIGHT 268#, BP 132/100, HEMOGLOBIN 16, HEMATOCRIT 50, RBC 5.35 MIL., WBC 12,000, VC 3 LITERS, ECG NORMAL. HE WAS TREATED WITH A 1000 CALORY DIET. BY 1951, WEIGHT WAS 345# AND EXERTIONAL DYSPNEA PROGRESSIVE. DIGITALIZATION IN 1952 BY HIS LOCAL DOCTOR WAS NOT PARTICULARLY BENEFICIAL, BUT HAS BEEN CONTINUED. BY 1953, VP 16, CT 12, ECG SHOWED RIGHT VENTRICULAR STRAIN, AND ARTERIAL 02 SATURATION OF 95% NOTED TO INCREASE TO 107% WITH 100% 02 BREATHING. IN 1955, VP 22.8, CT 28, BLOOD VOLUME 8 LITERS AND HE UNDERWENT CARDIAC CATHETERIZATION. AT THIS TIME, HE DESCRIBED CHRONIC COUGH, INCREASING RESPIRATORY DISTRESS TO THE POINT OF DYSPNEA AT REST WITH CYANOSIS AND EDEMA. SOON AFTERWARD HE WAS HOSPITALIZED FOR 8 MONTHS FOR DETAILED PULMONARY FUNCTION STUDIES, CONTROLLED DIETARY REGIME, AND PSYCHIATRIC EVALUATION AND THERAPY. IN ADDITION TO 58# WEIGHT LOSS (308# TO 250#), HE OBTAINED MARKED SYMPTOMATIC IMPROVEMENT, BUT HAS HAD SEVERAL SUBSEQUENT ADMISSIONS FOR CHRONIC COUGH, DYSPNEA, ORTHOPNEA, WHEEZING, PND, AND EDEMA. IN [redacted], 1958, HEMOGLOBIN WAS 17.8, HEMATOCRIT 58, CO_2 28, VP 21, CT 41, WEIGHT 232#, V.C. 2.6 LITERS AND 0.5 SEC. EC 1.2 LITERS HE WAS NOW CLEARLY SUFFERING FROM THE EFFECTS OF PROGRESSIVE OBSTRUCTIVE BRONCHOPULMONARY DISEASE IN ADDITION TO MECHANICALLY AND CENTRALLY INDUCED HYPOVENTILATION.

CASE 4. [redacted] A 58-YEAR-OLD WHITE MALE [redacted], HAD A LONG-STANDING HISTORY OF CHRONIC RECURRENT BRONCHOPULMONARY INFECTIONS OF A MILD TO MODERATE SEVERITY. HE SMOKED 1 1/2 TO 2 PACKAGES OF CIGARETTES PER DAY FOR 35 YEARS.

HE ENTERED THE HOSPITAL WITH A 4-DAY HISTORY OF EXACERBATION OF HIS ILLNESS, CHARACTERIZED BY INCREASING DYSPNEA, COUGH, MUCOPURULENT SPUTUM, AND LOW GRADE FEVER.

EXAMINATION REVEALED A SEMI-STUPOROUS, ACUTELY AND CHRONICALLY ILL MAN. BP 160/90, R. 32, P. 110. THE LUNGS REVEALED COARSE RHONCHI AND MEDIUM CRACKLING RALES WITH EXPIRATORY WHEEZES THROUGHOUT BOTH LUNGS. THE DISTRIBUTION OF THE FINDINGS VARIED SOMEWHAT WITH DEEP BREATHING AND COUGHING.

WBC 12,600, HEMOGLOBIN 17.0, HEMATOCRIT 60, BUN 43, CO_2 COMBINING POWER 48, CL 70, NA 133, K 4.0. OXYGEN SATURATION 48% ON ROOM AIR, pH 7.21. AFTER A SHORT PERIOD OF BREATHING 85% OXYGEN BY MASK, THE OXYGEN SATURATION WAS 94% BUT CO_2 COMBINING POWER WAS 52 AND pH 7.01. ECG REVEALED RIGHT VENTRICULAR HYPERTROPHY, AND X-RAY SHOWED MARKED BULLOUS EMPHYSEMA.

THE PATIENT WAS PLACED IN A TANK RESPIRATOR AND GIVEN BRONCHODILATOR NEBULIZATION BY IPPB Q 3 H. AMINOPHYLLINE AND PENICILLIN WERE GIVEN IN IV SOLUTIONS CONSISTING OF 2000 CC. R-L SOLUTION, 2000 CC. 5% DEXTROSE IN DISTILLED WATER.

THE NEXT DAY PHYSIOLOGICAL STUDIES WERE PERFORMED TO EVALUATE THE EFFECT OF ASSISTED RESPIRATION. (SEE TABLE). THE SAME TREATMENT REGIMEN WAS CONTINUED FOR THE NEXT 3 MONTHS WITH PATIENT SPENDING MOST OF EACH 2 1/2 HOUR PERIOD IN THE RESPIRATOR. SEVEN PHLEBOTOMIES (200 CC. EACH) WERE DONE DURING THIS PERIOD.

IMPROVEMENT WAS SLOW, BUT SUFFICIENT TO DISCONTINUE THE TANK AND RELY ON INTERMITTENT USE OF IPPB. AT THE TIME OF DISCHARGE HEMOGLOBIN WAS 13.6, HEMATOCRIT 47, BUN 15, CO2 COMBINING POWER 37, CL 94, OXYGEN SATURATION 78%, AND PH 7.42.

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Pulmonary Function Studies

W.V.H., 37 yr. WM

Date	1953	1955	1956
Weight	267#	294#	250#
0.5 Sec. EC	1.75	1.50	1.80
1.0 Sec. EC	2.2	1.70	2.3
VC	2.75	1.95	2.7
O2 Saturation	95	92	94
pCO_2 - room air	40	50	44
pCO_2 - 100% O2	48	60	49
pCO_2 - 100% O2 & IPPB - 10 minutes			33
pCO_2 - 100% O2 during MBC		38	

October, 1955 - Sitting and Supine Study

	Sitting	Supine	Predicted
V.C.	2.6 L.	2.1 L.	4.45
Functional Residual Capacity	2.5 L.	1.8 L.	--
Residual Volume	1.7 L.	1.6 L.	1.66
Total Capacity	4.3 L.	3.7 L.	5.55
RV/TC	40%	42%	30%
Index of Air Mixing	1.5%	7.0%	1.5%

B.McM., 34 yr. WM

5-8-57	Room Air Very Quiet	Room Air Forced Vent.	100% O2 Forced Vent.	100% O2 Quiet Breathing
O2 Saturation (%)	79.95	93.50	89.63	100
pCO_2 (mm. Hg)	64.0	51.5	50.0	58.0
pO_2 (polarograph) (mm. Hg)	48.5	78.0	57.0	483
pH at 37.5°	7.35	7.42	7.45	7.41
Transfer Gradient (Alv. pO_2-Art. pO_2)	20	0	92	171
Ventilation (L./min.)	3.25	4.1	4.8	3.8
Tidal Volume (cc.)	232	293	368	268

Pulmonary Function Studies
(continued)

B.McM.
5-14-58

	Room Air	2.32% CO_2	5.44% CO_2	100% O_2
O_2 Saturation	95.66	95.15	95.02	100
pCO_2 (mm. Hg)	36.5	50.5	66.0	58.0
pO_2 (polarograph) (mm. Hg)	99.0	98.0	106.0	491
pH at 37.5°	7.40	7.29	7.20	7.22
Transfer gradient	21	19	18	147
Ventilation (L/min) BTPS	7.91	8.15	11.13	6.54
Tidal Volume (cc.)	395	408	586	467

Physiological Studies

L.B.

	Room Air Unassisted	After 12 hr. IPPB-50% O_2	15 min. later Room Air Unassisted	30 min. later IPPB-Room Air	30 min. In sack Resp.	10 min. 85% O_2 Unassiste
CO_2 Content	36.54	26.85	26.91	26.65	26.36	30.60
O_2 Sat'n	80	100	88	91	91	100
pCO_2	88	58	62	55	50	71
pH	7.28	7.35	7.34	7.38	7.42	7.31
pO_2	53	274	63	65	63	450

H.H.

	Room Air	100% O_2 Unassisted	Tank Resp. Room Air	Tank Resp. 100% O_2	IPPB/1 After B.D.-100% O_2
O_2 Satn.	51	92	81	98	100
pCO_2	78	90	56	48	44
pH	7.31	7.28	7.40	7.41	7.43

OBESITY

EXCURSION OF THORAX AND DIAPHRAGM

ALVEOLAR HYPOVENTILATION

ALVEOLAR pO_2

ARTERIAL O_2 SATURATION

CYANOSIS
HYPERVOLEMIA
POLYCYTHEMIA
VISCOSITY

CARDIAC OUTPUT

PULMONARY HYPERTENSION

RIGHT-VENTRICLE HYPERTROPHY

RIGHT-VENTRICLE FAILURE

ALVEOLAR pCO_2

ARTERIAL CO_2

PULMONARY VASCULAR SPASM & SCLEROSIS

SOMNOLENCE
MENTAL CONFUSION
WEAKNESS
LASSITUDE
IRRITABILITY
TREMOR
TWITCHING
WARM, FLUSHING SKIN
SWEATING
TACHYCARDIA
HYPOTENSION
SENSITIVITY TO SMAL
CHANGES IN ACIDOSI

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