Addendum

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Dr. Donald R. McCurley of New Orleans has recently told me of studying and treating a patient with both "primary pulmonary hypertension" and adrenocortical insufficiency. The possible relationship of his observations to the findings described in the preceding report are quite apparent, and he has kindly permitted me to describe his case.

A 47 year old married woman with no children complained of fainting spells, fatigue, progressively increasing edema and exertional dyspnea. Prior to the increasing edema she had lost about 40 pounds (130 lbs. to 90 lbs.) in one year, despite a fair appetite. There was no past history of heart disease, murmurs, or rheumatic fever.

Admitting blood pressure was 170/104 but subsequent examinations demonstrated a labile blood pressure which was usually normal. There was brawny pigmentation in areas of dependent edema, but not in the skin creases nor the oral mucosa. Except for rales at both bases, and dependent edema, the remaining significant findings on physical examination were in the heart. The rhythm was regular at 62/min. and cardiac size seemed normal. At the apex there was a grade 2 systolic murmur, poorly transmitted, and also a diastolic rumble with accentuated first mitral sound. The second pulmonic sound was loud and split, and was followed by a blowing diastolic murmur transmitted down the left sternal border.

An electrocardiogram revealed marked right ventricular hypertrophy, with a qR in V1 (R of 10 mm.) and an rS (predominantly S) in V6. On x-ray examination the right ventricle and main pulmonary artery segment were markedly enlarged, with equivocal enlargement of the left ventricle and atrium; the right atrium was moderately enlarged.

At cardiac catheterization a coiled catheter demonstrated a huge right atrium. Pressures in the right ventricle, main pulmonary artery and pulmonary wedge position were, respectively (in mm. Hg), 132/10, 132/55, and 10 (mean). All blood oxygen examinations were normal, and there was no evidence of shunt from either hydrogen-platinum electrode nor cardiogreen dye studies. At the end of the procedure the patient developed paroxysms of nodal premature beats. During an angiocardiogram the next day she became hypotensive (70/40 mm. Hg) but responded promptly to intravenous Wyamine. The angiocardiograms demonstrated a giant right atrium. The final diagnosis was primary pulmonary hypertension with a giant right atrium.

Because of suspected adrenocortical insufficiency, urinary steroid excretion studies were done. Control levels were 2.3 mg. of 17 ketosteroids and 7.0 mg. of 17 hydroxycorticosteroids excretion in 24 hours. The next two days 60 units of ACTH gel was injected intramuscularly each day and urine collected for each of the subsequent 24 hours. Volume of urine the first day was 1120 ml. with 4.2 mg. of 17 ketosteroids and 15.0 mg. of 17 hydroxycorticosteroids. Volume of urine the second day was 1820 ml. with 8.2 mg. of 17 ketosteroids and 26.0 mg. of 17 hydroxycorticosteroids. A diagnosis of moderate adrenocortical insufficiency was made.

Pulmonary Hypertension

On her discharge from the hospital the digitalis which she had been receiving was discontinued, and 5 mg. of preonisone twice daily was prescribed. On this treatment her appetite improved, she gained 8 lbs. in one month, and had no further edema. Exercise tolerance improved.

If it can be presumed that this patient's adrenal medullary secretions failed at the same time as those from the adrenal cortex, then the postulated contributing role of catecholamines to the pathology of DAPH suggested in the preceding report is strengthened. It would of course be premature to make sweeping conclusions from these preliminary observations, but the clinical events and course of this unusual case fit so well with the morphologic lesions described (especially Case 2) that it was felt they merited description. Dr. McCurley's patient presents but another reason adrenal medullary and cortical function may require more careful evaluation in future cases of degenerative arteriopathy with pulmonary hypertension.