A CASE OF PECULIAR CUTANEOUS PIGMENTATION, PROBABLY AN INCOMPLETE FORM OF RECKLINGHAUSEN'S DISEASE.

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The patient is a fairly well-developed girl, aged 15 years. The pigmentation affects the trunk and neck chiefly, and the extremities to a much lesser degree, the face being almost free. There are no areas of leucoderma. There is no evidence of urticaria, factitious urticaria, purpura, or any kind of erythema in connection with the condition. The patient presents, roughly speaking, three different kinds of cutaneous pigmentation, namely: (1) Diffuse brownish patches, especially a large patch over the upper part of the back and neck, which has a sharply defined, probably stationary, upper border, but is ill-defined below, in which direction it is apparently spreading and merges gradually into the ordinary skin. (2) Brown spots and small patches plentifully scattered over the trunk, the paler ones being probably those most recently developed. (3) A group of very dark, almost black, spots on the left side of the thorax, resembling a group of “pigment naevi,” but not raised above the general level of the skin. These blackish spots were first noticed about three years ago. The earliest pigmentation noticed by the mother was at the back of the neck when the child was only about eighteen months old. Since that time the rest of the pigmentation has gradually developed, and fresh, relatively faint spots and patches have lately appeared on the extremities. The patient’s general nutrition is fairly good, and menstruation has recently commenced. Though she looks rather pale, examination of the blood (Dr. Schenck) has shown nothing of pathological significance. The brachial blood-pressure by the Riva-Rocci instrument was found to be about 130 mm. mercury. The examination of the thoracic and abdominal viscera and of the urine shows nothing abnormal. Cutaneous sensation, knee-jerks, and plantar reflexes are natural. The mental develop-

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ment and general intelligence are up to the average. There is no pigmentation in the mucous membrane of the mouth. In the past

history of the patient the only noteworthy point is that she has always been very liable to headache and bilious vomiting. But
there has been no tendency to fainting such as to suggest Addison's disease. On the contrary, the attacks of vomiting seem to have been induced by certain articles of food, such as rich, fatty things and, at one time, raw apples. Moreover, the patient's mother, as a child and young woman, used to be subject to so-called "bilious attacks" (it may be noted in this connection that the mother has small patches of Xanthelasma palpebrarum, which appeared some years ago), and one or two others in the family have been inclined to similar attacks. Apparently no anomalous pigmentation has been observed in the family except in the patient herself. In her case, from the diagnostic point of view, it is to be noted that the pigmentation affects chiefly the covered parts of the body, and that there is no special liability to freckling from exposure to the sun. This is the reverse of what occurs in cases of Kaposi's Xeroderma pigmentosa. The variety in the colour and form of the areas of pigmentation and their situation (mainly on the trunk) make it seem most probable that the present case is allied to (that is to say, a forme fruste, or incomplete form of) Recklinghausen's disease—in fact, that it is a case of neuro-fibromatosis, with typical cutaneous pigmentation, but, as yet, practically without any (superficial) tumours. There is only a single small, flaccid, molluscous tumour to be found. This is situated on the lower part of the back and was first noticed about three years ago. Probably no other satisfactory explanation for the pigmentation can be offered except that mentioned above, which was first suggested by Dr. A. Whitfield when he was told about the case.