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alert child who is likely to outstrip his fellows in the battle of life, provided he is not hampered by disabilities resulting from such reactions as eczema." Questions 3, 4, 5, and 6 were asked in an attempt to support or refute such statements. Failure to do either was certain unless the figures argued strongly one way. They did not, and therefore any method short of thorough psychological examination of each case cannot help in the matter. It is interesting to note that the only males confessing inadequacy of emotional make-up were those afflicted by asthma or Besnier's prurigo, or both.

The opinion is often quoted that the scalp is not usually affected in cases of infantile eczema which are destined to progress to Besnier's prurigo. In this series 12 of those with Besnier's prurigo had had scalp involvement as infants and 11 had not. However, almost 90% of the cases of Besnier's prurigo had had infantile eczema on the trunk and limbs (as well as on the face and/or scalp).

Conclusions

When the figures for hay-fever only, asthma only, Besnier's prurigo only, and both asthma and Besnier's prurigo are added together it is found that out of the 93 useful replies received, 63 describe at least one of the recognized sequelae of infantile eczema. Of these 63, 52 have had sequelae of no minor severity. Thus we find that, of the group of infants with eczema under study, between 33 and 56% were destined to many years or a lifetime of grave disability. This, when one is confronted with a case of infantile eczema, is a sobering thought. May it stimulate the continued untiring investigation of the problem.

Considerations of the restricted locale, the moderate numbers, the isolated period, and the wide range of percentages, which are all unavoidable components of this investigation, preclude generalization.

Sneddon (1951), in an excellent contribution, has reemphasized the effect upon prognosis of a strong family history of eczema and asthma, the psychological background of the family, and over-protection. In an article which attempts to educe actual figures these undeniably important influences are relatively imponderable and have therefore not been considered.

It is hoped that others will find opportunity, in different centres, to follow up the cases of infantile eczema of years ago, so that eventually the figures will be large enough to give a high degree of accuracy.

Summary

An investigation of the incidence of sequelae following infantile eczema of fifteen to twenty-one years ago is described.

Between one-third and one-half of the cases studied have sequelae sufficient to encumber at least their first fifteen to twenty years of life.

Spasmodic asthma leads the field, affecting between one-quarter and one-half of the cases.

Besnier's prurigo has afflicted roughly one-fifth of

Allergic rhinitis is found in one-tenth.

I wish to record my gratitude to Professor G. H. Percival and Dr. Robert Aitken for permission to use their cases and for their advice and encouragement. I wish also to thank Dr. G. A. Grant Peterkin for suggesting the investigation in the first instance and for his subsequent invaluable help and criticism.

REFERENCES

Abt, I. A. (1909). J. Amer. med. Ass., 53, 840.

Bamber, G. W. (1931). Brit. J. Derm., 43, 295.

Besnier, E. (1892). Ann. Derm. Syph., Paris, 3rd ser., 3, 634. Quoted by Bray (1937, p. 340).

Bray, G. W. (1937). Recent Advances in Allergy, 3rd ed., p. 199. Churchill, London.

Derbes, V. J., and Engelhardt, H. T. (1943). Amer. J. med. Sci., 205, 676.

Ellis, D. (1933). Guy's Hosp. Rep., 83, 102.

Hall, A. J. (1908). Brit. J. Derm.. 20, 8.

Hill, L. W. (1942). J. Pediat., 20, 537.
— and Sulzberger, M. B. (1935). Arch. Derm. Syph., Chicago, 32, 455. Kurtz, M. (1950). N.Y. St. J. Med., 50, 1247.
McGregor, R. M. (1950). Edinb. med. J., 57, 433.
Rasch, C. (1914). Nord. med. Ark., 2, 9. Quoted by Bray (1937, p. 341). Sequeira, J. H., Ingram, J. T., and Brain, R. T. (1947). Diseases of the Skin, 5th ed., p. 153. Churchill, London. Sneddon, I. B. (1951). Med. Press, 226, 329.
Taylor, R. W. (1899). N.Y. med. J., 70, 593. Quoted by Bray (1937, p. 335).

A CASE OF MALE **PSEUDOHERMAPHRODITISM**

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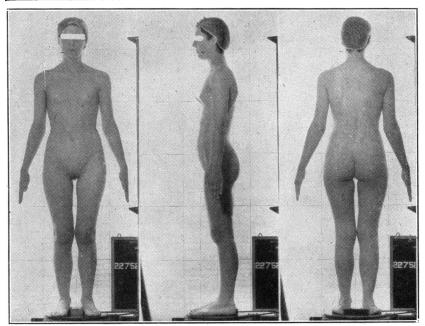
Schneider, Van Ommen, and Hoerr (1952) have refocused attention upon the condition of male pseudohermaphroditism. Williams (1952) refers to the condition as "intersex males with purely feminine external genitalia and bodily habits." These patients have a normal feminine mentality and normal or greater than normal libido. Breast development is normal, though the nipples and areolae tend to be small. The external genitalia are normal, but the labia are small. Axillary hair is absent and the pubic hair scanty or absent. There is a short vaginal pouch which ends blindly, the cervix is absent, and the uterus is absent or rudimentary. The gonad is usually intra-abdominal, most often in the position of the ovary, but not infrequently it is in the inguinal canal, when it may be associated with a hernia. A moderate degree of vaginal cornification is present and the 17-ketosteroid output is at the normal feminine level. There is often a familial incidence.

Schneider et al. laid down definite criteria for the These are: feminine habitus, primary amenorrhoea, absence or almost complete absence of axillary and pubic hair, blind vaginal pouch with absent cervix, and intra-abdominal testis. They could find only 12 cases in the literature which fulfilled their criteria, and reported six cases of their own. We would like to report a further case which was diagnosed clinically before the result of the previous operation was known.

Case Report

A single woman aged 25 was admitted on May 7, 1952, complaining of epigastric pain half an hour after meals and of backache of three weeks' duration. She had not vomited and her appetite was good. She had lost 14 lb. (6.4 kg.) in weight during this time. Recently she had become paler and had noticed that the stools were dark. She had a dry cough since the age of 18 which she attributed to smoking.

She had never menstruated. Breast development and some pubic hair appeared at the age of 16. At the age of 17 she was 5 ft. 2 in. (157 cm.) in height and weighed 9 st. (57.2 kg.). From then until the age of 21 she grew rapidly to 5 ft. 8½ in. (174 cm.), when she weighed 9 st. 7 lb. (60.3 kg.). Growth then ceased. She has a normal feminine outlook and was recently engaged to be married. engagement was broken off by her fiancé because he found her sexually unresponsive. This led to her becoming somewhat introspective. There are no homosexual inclinations.



Photographs of the patient

Past History.—She had measles and mumps as a child. At the age of 5 (in 1932) an operation was performed at Newcastle Royal Infirmary for left inguinal hernia. At the age of 18 she was again seen at that hospital. An examination under anaesthesia was carried out and the uterus was found to be absent.

Family History.—Her father died eight years ago from a cerebral haemorrhage. Her mother is alive and well, apart from indigestion. One uncle died from a stroke, one suffers from indigestion, and another is in Australia. One aunt died aged 26 years (cause unknown) and the other suffers from Bright's disease. She has one brother, who is married. One sister died from meningitis at the age of 5. Three other sisters are alive and well, married, and have families. Their periods did not begin until the age of 17 or 18. Pubertal development occurred between the ages of 13 and 14.

On admission the patient's height was 5 ft. $8\frac{1}{2}$ in. (174 cm.); weight 8 st. 10 lb. (55.3 kg.); vertex to symphysis, 35 in. (89 cm.); symphysis to ground, $33\frac{1}{2}$ in. (85 cm.); span of outstretched arms, 69 in. (175 cm.); feminine habitus. The voice was rather low-pitched and husky. The mucous membranes were pale. Numerous freckles were present over the face and arms. Axillary hair was absent; pubic hair fine and scanty. The nipples and areolae were small. The breasts were small, but mammary tissue was present. The external genitalia were normal, though the labia were small. Cardiovascular system: blood pressure, 130/70; heart sounds normal. Respiratory system: normal. Abdomen: tender in the epigastrium, but no mass or viscus felt. There was a scar from a left inguinal hernia operation; no herniae present. Central nervous system: retinal veins somewhat tortuous and slight filling of the optic cup; there was no other abnormality. Rectal examination: no uterus or cervix was felt.

Investigations.—Blood count: red cells, 4,400,000; haemoglobin, 9.2 g.% (62%); colour index, 0.7; white cells, 12,500 (polymorphs 65%, lymphocytes 32%, monocytes 3%). Thorn's test—eosinophil count before A.C.T.H., 60 per c.mm.; four hours afterwards, 40 per c.mm. B.M.R., +5%. Plasma cholesterol, 150 mg. per 100 ml. Fractional test meal, normal acid curve. Occult blood, positive. Barium meal: there is a large ulcer crater on the posterior wall in the pars media and a small one just above the first. The duodenal cap is normal. Glucose-tolerance test (100 g.): fasting, 81 mg. per 100 ml.; after ½ hour, 121 mg.; 1 hour, 144 mg.; 1½ hours, 126 mg.; 2 hours, 126 mg. No glycosuria. Skull: no abnormality seen; sella turcica

of normal appearance. Wrists and knee-joints: epiphyses have fused. Output of 17-ketosteroids, 9.2 mg. in 24 hours. Urinary gonadotrophin output, less than 10 units in 24 hours. Urinary oestrogens: oestrone, 1.5 μ g. in 24 hours; oestradiol, 2.5 μ g.; oestroil, 2.0 μ g. Vaginal smear: moderate cornification (++ to +++).

Discussion

On clinical examination this patient seemed to fulfil the criteria of Schneider et al. She was tall and slim, quite unlike the short stocky build resulting from ovarian agenesis. Furthermore, breast development was present. Ovarian agenesis was also excluded by the degree of vaginal cornification present and because the output of urinary gonadotrophin was not increased. Her height, her freckled appearance, and the presence of some pubic hair were equally unlike a primary pituitary dysfunction. This was confirmed by the normal basal metabolic rate, glucosetolerance curve, and urinary output of 17-ketosteroids.

Exploratory laparotomy was contemplated to confirm the diagnosis. This proved unnecessary, however, as Professor Duguid kindly lent us the histological sections of the gonad removed at the operation for inguinal hernia when the patient was 5 years old. This showed the typical appearances of a cryptorchid testis.

The diagnosis of male pseudohermaphroditism should be considered whenever a patient with primary amenorrhoea is found to have absent axillary hair and absent or scanty pubic hair with nearly normal breast development and normal external genitalia, and in whom an examination under anaesthesia shows the uterus to be absent. Although there was no family history in our case, the condition is often familial. The presence of an inguinal hernia or the scar of a previous operation for the repair of one is common in this condition. The vaginal smear provides a simple means of demonstrating some degree of oestrogenic activity and of thereby excluding primary ovarian failure, as in ovarian agenesis, as the cause of the amenorrhoea. The diagnosis can generally be confirmed only by laparotomy and gonadal biopsy; it is probable that cases have been missed through the assumption that the gonad found on laparotomy was an ovary. As the testis is the source of oestrogens, castration is contraindicated: Williams described a case in which castration was followed by the development of menopausal symptoms and genital atrophy.

Summary

A case of male pseudohermaphroditism is presented. The clinical features of this condition are reviewed and suggestions made regarding methods of diagnosis.

Our thanks are due to Dr. Una Ledingham and Miss J. Moore for permission to publish the case, to Professor Duguid for kindly lending us the histological section, and to Dr. J. M. Tanner for the photographs of the patient.

REFERENCES

Schneider, R. W., Van Ommen, R. A., and Hoerr, S. O. (1952). J. elin. Endocr., 12, 423.
Williams, D. I. (1952). British Medical Journal, 1, 1264.

Among those included in the Coronation Honours List was Sister Catherine Dynes, who was awarded the M.B.E. Sister Dynes has been Night Superintendent in charge of the Royal Victoria Hospital, Belfast, since 1909, and her award will be welcomed by a wide circle of Belfast medical men.