A GIRL OF MONGOLOID APPEARANCE AND NORMAL INTELLIGENCE.

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During a domiciliary visit one of us encountered a child who appeared to be a typical mongol and who nevertheless behaved remarkably intelligently. This child was later shown to numerous colleagues, among them the late Sir Leonard Parsons, who spontaneously suggested that she was a "mongol." The combination of mongoloid features, which she exhibits, and normal intelligence is of sufficient interest, we feel, to be recorded.

PHYSICAL APPEARANCE.

The child in question is now fourteen years old, a brunette of average height (59\frac{1}{2} in.) and weight (84 lb.). Her head is small (circumference 19\frac{1}{2} in.), bulletshaped and brachycephalic with a low cephalic index .875 (anterior posterior diameter 6.3 in. (16 cm.), lateral diameter 5.5 in. (14 cm.)). Her eyes are widely separated and almond-shaped; the palpebral fissures are narrow and slope downwards and inwards (Fig. 1); she has an oblique hypermetropic astigmatism and a concomitant strabismus. On one occasion she was admitted to hospital for a left graded tenotomy, but operation was deferred when it was discovered that she had congenital heart disease; she has in fact a patent ductus arteriosus. Her irides are brown and, naturally (Wallis, 1951), no Brushfield's spots are present. Her tongue is of normal size with a deep central furrow; it is not repeatedly sucked in and out and has therefore not a hacked or scrotal appearance (Thomson, 1898). Her ears are small, have a simple design and no lobe. The little finger of each hand is curved and shortened (Fig. 2) with a short middle metacarpal; three main flexures cross each palm; the dermal pattern is not typical of mongolism (Cummins, 1939), but it differs radically from the pattern on the palms of her parents and siblings, and her ridge count, an important feature (Fang, 1949), is also noticeably less. Her feet and toes are normal.

This child is in the top class at school, she has a good record and is soon to enter a secretarial college. Her intelligence quotient (Terman Merrill) is 116.

PAST HISTORY.

She was born prematurely (birth weight 4 lb.) after an uneventful pregnancy, of young, unrelated parents; the father was 23 and the mother 24 years old at her birth. She is the oldest child in her family. There have been five subsequent pregnancies, resulting in one stillbirth and four other children,



Fig. 1.

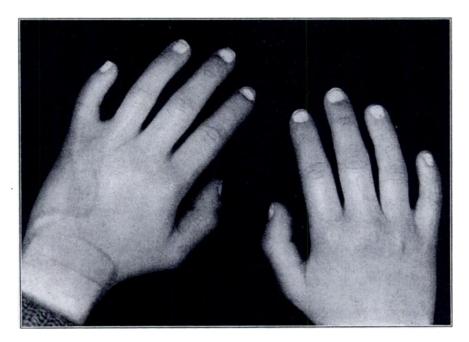


FIG. 2.

none of whom has a mongoloid appearance. The girl's dentition was normal, but her head, according to her grandparents, was very late in closing. She sat up at eight months, walked at fifteen months, said odd words at ten months and could read simple sentences at the age of 5 years.

Discussion.

The concept of mongolian idiocy, originally put forward by Langdon Down (1866) was based on the frequent association, in certain oligophrenics, of obvious mental retardation and mongolian features. Although Langdon Down himself did not fully analyse the individual features which together build up the mongol's appearance, this has been done by subsequent writers, who have also drawn attention to the frequency with which other abnormalities, e.g., congenital heart disease and cataract, occur in mongols. The main physical characteristics of mongols are:

- 1. Dwarfing.
- 2. Brachycephaly.
- 3. Widely separated, obliquely inclined almond eyes.
- 4. Narrow palpebral fissures and epicanthic folds.
- 5. Errors of refraction (hypermetropic astigmatism), strabismus, cataract and Brushfield's spots.
- 6. A hacked or scrotal tongue.
- 7. Congenital heart disease.
- 8. A small curved little finger.
- 9. A certain dermatoglyphic pattern on the palm of the hand.
- 10. Widely spaced first and second toes with a deep furrow on the sole of the foot.
- 11. Hyperextensibility or laxity of joints.

In addition the mongol is often the product of elderly parents, born towards the end of the mother's productive years.

The diagnosis of mongolism in any individual subject is not dependent on the simultaneous presence of all these features; it is based rather on the subject's general appearance, i.e., on his likeness to a mongol (Langdon Down, 1866); no single feature taken by itself either justifies or is essential to the diagnosis (Weber, 1917), for individual mongolian characteristics are quite common in oliophrenics (Penrose, 1933), and they are also not rare in normals. Only when several of these features occur in the same person can the diagnosis of mongolism be justified (Tredgold, 1947), and in these, according to Tredgold, there will usually be found some pecularity of the skull, eyes, tongue and hands; peculiarities of all these are present in our case; in addition she has congenital heart disease, and of a type not uncommon in mongols (Tredgold, 1947).

Sutherland (1907) demonstrated a girl of eight who had a shortened curved little finger and obliquely slanting eyes—a typical mongol apparently—who was also intelligent, and it would seem that mongolism need not necessarily be associated with either idiocy or imbecility (Weber, 1917). The child of whom we have given details, like Sutherland's case, also has the appearance of

a mongol and is nevertheless intelligent; she differs, however, from most mongols in being of normal stature, and from some in being the oldest child in the family and in being born of young parents, but the latter is also true of many other mongols (Still, 1898); Tredgold (1947) even gives one instance of a mongol being born to a girl of 16. This case is recorded because the association of so many mongolian features and normal intelligence is unusual; we would also like to emphasize that mongolian stigmata are not always associated with mental retardation.

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