ILLEGITIMACY AND TWINNING IN PATIENTS WITH C.N.S. DEFECTS

—	S.B.	C.D.	H.C.	M.C.
No. legitimate patients ..	35	15	44	59
No. illegitimate patients ..	5	3	12	9
Total patients..	40	18	56	68
% Illegitimate patients ..	12·5	16·7	21·4	13·2
Mean parity	2·88	3·00	2·62	2·73
Mean sibship size*	3·94	4·27	4·39	4·05
Sex ratio of patients	(18/22) 0·82	(10/8) 1·25	(35/21) 1·67	(23/44) 0·51
Sex ratio of liveborn sibs* ..	(56/47) 1·19	(30/19) 1·58	(78/65) 1·20	(74/103) 0·72
Twinning in patients ..	(1/40) 2·5%	(1/18) 5·6%	(1/56) 1·8%	(5/68) 7·5%
χ^2; (P)	0·755	3·417	0·263	26·69 (0·001)
Twinning in liveborn sibs* ..	(4/98) 4·1%	(1/48) 2·0%	(1/148) 0·7%	(0/180) 0%
χ^2; (P)	8·283 (0·01)	0·453	0·226	2·561

* For legitimate patients only. No recent attempts made to determine completeness of sibships.

twin patients in these families, 1 preceded the birth of dizygotic twins, 1 followed the birth of dizygotic twins, and 1 followed the birth of male twins, zygosity unknown. The mothers were 30, 28, and 28 years of age respectively. The incidence of twinning in Wisconsin is 1·08%. The incidences among both the M.C. patients and the sibs of the S.B. patients are significantly higher than this.

1 of the 3 S.B. patients is a twin; his twin sister is hydrocephalic. His mother's brother has two hydrocephalic children, a boy and a girl. Whether this case represents an example of fetus-fetus interaction [11–13] or a coincidental occurrence of a segregating trait cannot be determined. 1 C.D. patient, 1 of 2 H.C. patients, and all 5 M.C. patients are twins. The co-twin of the C.D. patient had been dead about two days at birth. Both twins were male, but no other data on zygosity are available. All 5 M.C. twins are female and all 5 co-twins are abnormal. In 1 case, the co-twin is an epileptic male, and in 1 it was a stillborn anencephalic, sex and zygosity unknown. 1 co-twin was "incompletely formed". The co-twin of 2 patients died during the fourth or fifth month of gestation; 1 of these was monochorionic and diamniotic with a velamentous insertion on an infarcted area of the common placenta: no information is available on the other. The C.D. twin was not thought to be defective at birth, but the M.C. twins were immediately recognised as microcephalic. It is possible that the presence of a dead co-twin may damage the developing C.N.S. and that the longer the exposure, the earlier and greater the damage to the surviving partner. The surviving twins were possibly normal until placental failure or abnormal development caused the death of their co-twins. This mechanism of fetus-fetus interaction may account for a proportion of C.N.S. defects in twin gestations. In any event, our data support the concept of fetus-fetus interaction in the M.C. group.

Our data support the hypothesis of an association between twinning and C.N.S. defects, not only for A.S.B. but for all types. Some M.C. and H.C. is obviously due to trauma in twin and non-twin deliveries, and necropsy may be necessary before a developmental defect is recognised. Familial forms of both H.C. and M.C. are known. In our study, twinning has *not* been found in families with segregating H.C. or M.C. However, among our 7 S.B. families with S.B. and/or H.C. in other family members, there are 2 families with twinning. Our data also suggest that after a dizygous twin delivery in which at least one partner has a gross C.N.S. defect, or after a monozygous twin delivery in which one twin is stillborn and the other is affected, the recurrence risk may be less than the recurrence risk (about 5%) usually cited for singletons for such a defect.

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INFLUENZA A IN AN ISOLATED POPULATION IN THE AMAZON

SIR,—We wish to report evidence of activity of H_2N_2 (Asian influenza A) in man two years after H_3N_2 strains became dominant around the world. The H_3N_2 epidemics of 1968–69 displaced the predecessor H_2N_2 strains so rapidly that within 12 months new isolations of the older strains had ceased.[1] Nevertheless, the discovery in pre-1957 sera of H_2N_2 antibodies in persons born before 1890,[2] and H_3N_2 antibodies in persons born before 1902,[3] suggests that these strains which became dominant in 1957 and 1968, respectively, were not truly novel. It is unknown whether the ascendancy of a new major strain represents emergence of old types from an active biological reservoir, or reconstitution from genetic components that had persisted, if at all, only in other combinations.

INFLUENZA A H-I ANTIBODIES IN MEKRANOTI SERA

Age in 1972	No. tested		H_2N_2 TW/64	HK/68	H_3N_2 Eng/72
<5	12	%+	83	0	0
		G.M.T.*	121	0	0
5–19	22	%+	100	23	30
		G.M.T.	106	20	56
>20	28	%+	100	31	63
		G.M.T.	122	47	37

* Geometric mean titre of positive specimens.

We have been studying very isolated Indian populations of the Brazilian Amazon and have found several groups serologically virgin with respect to influenza.[4] However, positive hæmagglutination-inhibition reactions were obtained with all but 2 of 61 serum specimens collected from the Mekranoti Indians in June, 1972, when Taiwan/64 H_2N_2 antigen was used (see accompanying table). These sera were negative with PR8/34 antigen and only a few low titres were obtained with the more recent H_3N_2 strains (HK/68 and Eng/72).

Ages of the members of this tribe could only be estimated, but H_2N_2-positive specimens came from all age-groups, including the youngest. Positive reactions were found in 4 children with only 16–20 teeth (estimated age 16–25

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months). The 2 negative specimens were from children with 20 and 22 teeth. This would suggest that the H_2N_2 virus had been active in this community during 1970–71, two years after it was displaced as the dominant worldwide strain.

We do not know whether the H_2N_2 virus had persisted in the Indian community because the other strains had never been introduced to displace it, or whether it had persisted at low frequency in the larger Brazilian community and had been introduced from there. Other evidence suggests that influenza would be unlikely to persist in a population as small as the Mekranoti (192 persons) for lack of an adequate supply of new susceptible persons.[4]

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MALIGNANT KWASHIORKOR

SIR,—The adjective "malignant" has been used to describe several conditions with a poor, or potentially poor, prognosis—malignant hypertension, malignant ophthalmos, malignant malaria, malignant melanoma, and so on. The disease kwashiorkor has also been termed "malignant malnutrition."[5]

Until the late 1940s the case mortality-rate of kwashiorkor was 80–100%.[6] After the recognition that protein deficiency was responsible for the condition the hospital mortality-rate fell rapidly but has remained fairly constantly at 20–50% for over twenty years.

It has long been recognised that there is a hard core of cases in which death comes suddenly within 24–48 hours of admission, and, as early as 1933, Cicely Williams noted that at least one clinical sign was associated with an early fatal outcome.[7] This sign was that of hæmorrhagic dermatosis (see accompanying figure, courtesy of Dr. F. M. Shattock).

Three biochemical tests and the accompanying electrocardiographic vector have also been found to have similar prognostic significance.[8] We therefore suggest that the term "malignant kwashiorkor" be given to those cases which have any of the following criteria: (i) acute hæmorrhagic dermatosis; (ii) limb-lead E.C.G. vector $\geq 60°$; (iii) total serum-protein values $\leq 3{\cdot}5$ g. per 100 ml.; (iv) serum-pseudocholinesterase values ≤ 10 units; (v) serum-siderophilin values ≤ 20 mg. per 100 ml.

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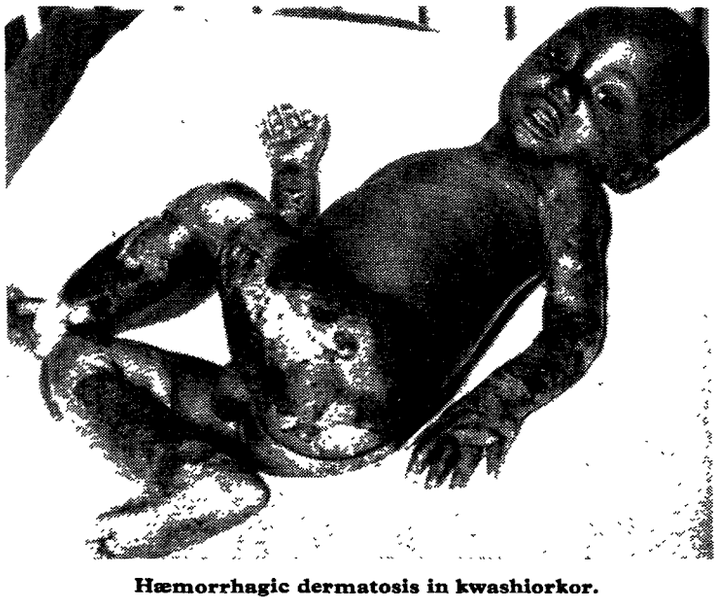

Hæmorrhagic dermatosis in kwashiorkor.

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MALNUTRITION AND SCHOOL PROGRESS

SIR,—Many reports have appeared on the adverse effects of protein energy malnutrition (P.E.M.) on intelligence. Inferior performance in intelligence tests in the affected has been well documented. However, there is no certainty over the precise degree of responsibility shared between malnutrition and other deleterious environmental factors. Understandably, many attach principal blame to malnutrition,[1,2] yet at a recent symposium [3] held in Sweden it was concluded that evidence "that malnutrition in early life jeopardises mental development . . . is scanty", and that intellectual status was the resultant of "the interaction between malnutrition and other environmental factors". Dr Dobbing (April 27, p. 802) emphasised that our chief concern should be with "a child's capacity for achievement" rather than with mental retardation per se, with its usually implied pathological connotation. He and others, particularly Evans et al.,[4] have stressed the multifactorial nature of achievement, and the difficulties entailed in its assessment.

In an attempt to throw light on the problem, a long-term follow-up study (details of which will be published elsewhere) is being carried out on the scholastic progress of previous P.E.M. sufferers, and also non-sufferers, in a rural South African Negro school. We wish to draw attention to two important observations: (1) children with a history of P.E.M. had twice the failure-rate in their school grades compared with control pupils; (2) among the affected group, learning difficulties were greater in younger than in older children. In younger children in primary grade, failures were approximately 40%, compared with about 20% in older groups in later grades; it was not unusual for younger children to repeat the first grade for three or four consecutive years. Clearly, a proportion of P.E.M. sufferers are displaying a "catch-up" in their learning ability after a variable period of adjustment to the school environment. This indicates that the early adverse situation, in the long run, may be less damaging than is at present supposed. Obviously, future studies must concentrate on (1) the very long-term effect on the sufferers, not only throughout school, but also in early adult life; (2) the decrease in the "capacity for achievement" amongst the far commoner mild and moderate forms of P.E.M., as emphasised at the symposium in Sweden. It must be reiterated that caution must be exercised in prediction. In long-term studies on South African Negro children with another type of disability, schistosomiasis, we were unable to detect significant differences in growth, blood biochemistry, or scholastic ability between the infected and non-infected.[5]

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