A CASE OF MONGOLISM IN DZ FEMALE TWINS STUDIED AT 10 AND THEN AT 43 YEARS OF AGE

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A pair of female mongoloid twins was studied when they were ten years old and, again, some thirty years later. A careful comparison was made of the similarities and differences between them in features or traits which have proved to be useful in determining the zygosity of like-sexed twins.
Though some of these characters can be, and frequently are, considerably modified in the presence of mongolism and so lose part of their value for that purpose, the differences noted in these twins, both as children and as middle-aged adults, are thought to outweigh their similarities and so warrant the conclusion that they are, indeed, dizygotic.

INTRODUCTION

More than 200 cases of mongolism (Down's syndrome) in twins have been reported. Thirty-seven of these are included in Table 3 of Brousseau's (1928) monograph on Mongolism (p. 37), which lists those which she had been able to find in the literature up to 1926.

The earliest of those cases is that attributed to Fraser and Mitchell (1876). The last half of their paper consists of notes by Dr. Mitchell in which he records numerous interesting observations that he had made on " Kalmuc Idiocy " over a period of many years. Mitchell ended his notes by listing what he described as " all the facts which I have recorded [on this subject] and which seem of any value." There are 25 of these " facts ", number 19 of which states merely: " In another [case], the idiot was one of twins." No mention is made of the sex of the twins or of the mental condition of the other member of the pair.

Thirty-six years after the publication of Brousseau's monograph, de Wolff et al. (1962, Table I) compiled 201 cases of mongolism among twins. In 169 of these, only one member of each pair was mongoloid. Of the 32 cases in which both twins were affected, 18 had been recorded as MZ; in 12 pairs the zygosity could not be conclusively determined; and only 2 pairs, one like-sexed and the other unlike-sexed, were considered to be DZ. More recently, McDonald (1964) reported on two pairs of unlike-sexed twins in which both members of each pair were mongoloid, and Fielding and Walker (1972) described a similar case of " dizygotic

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twins of opposite sex with unequivocal Down's syndrome substantiated by chromosome analyses."

The rarity of mongolism in both members of a DZ twin pair prompts me to record what appears to be another such case. It might prove to be of especial interest, because of the amount of available information from which the zygosity of the twins can be inferred and, also, because both twins were reexamined and additional relevant data obtained on them, more than thirty years after our first observations were made.

THE ORIGINAL FINDINGS

I first saw these twins at the Adolescence Study Unit of Yale University School of Medicine, in March 1939, through the kindness of Dr. Helen Thompson, who was then a member of the staff of Dr. Arnold Gesell's Clinic there. Photographs of the twins and roentgenograms of their skulls and hands which were made at that time are shown in Figs. 1 through 4.

The children were 10 years and 5 months old, when the photographs and roentgenograms were made. Twin A was then 6 cm taller than her sister (140.5 cm as compared with 134.3 cm). She was, also, longer-legged: the sitting-height/standing-height ratios of Twin A and B were, respectively, 54.5 and 55.2. As their photographs suggest, Twin A appeared to be somewhat less mongoloid than her sister.

The maximum head length of Twin A was 16.3 cm and her maximum head breadth was 13.7 cm. The corresponding dimensions of Twin B were 16.6 cm and 13.8 cm. The cephalic index of Twin A was 84.0 and that of Twin B was 83.1.

The lateral and the posterior-anterior roentgenograms of their skulls are reproduced in Figs. 2 and 3. In the original lateral skull film of Twin B, a few calcified granules are visible in the pineal

Fig. 1

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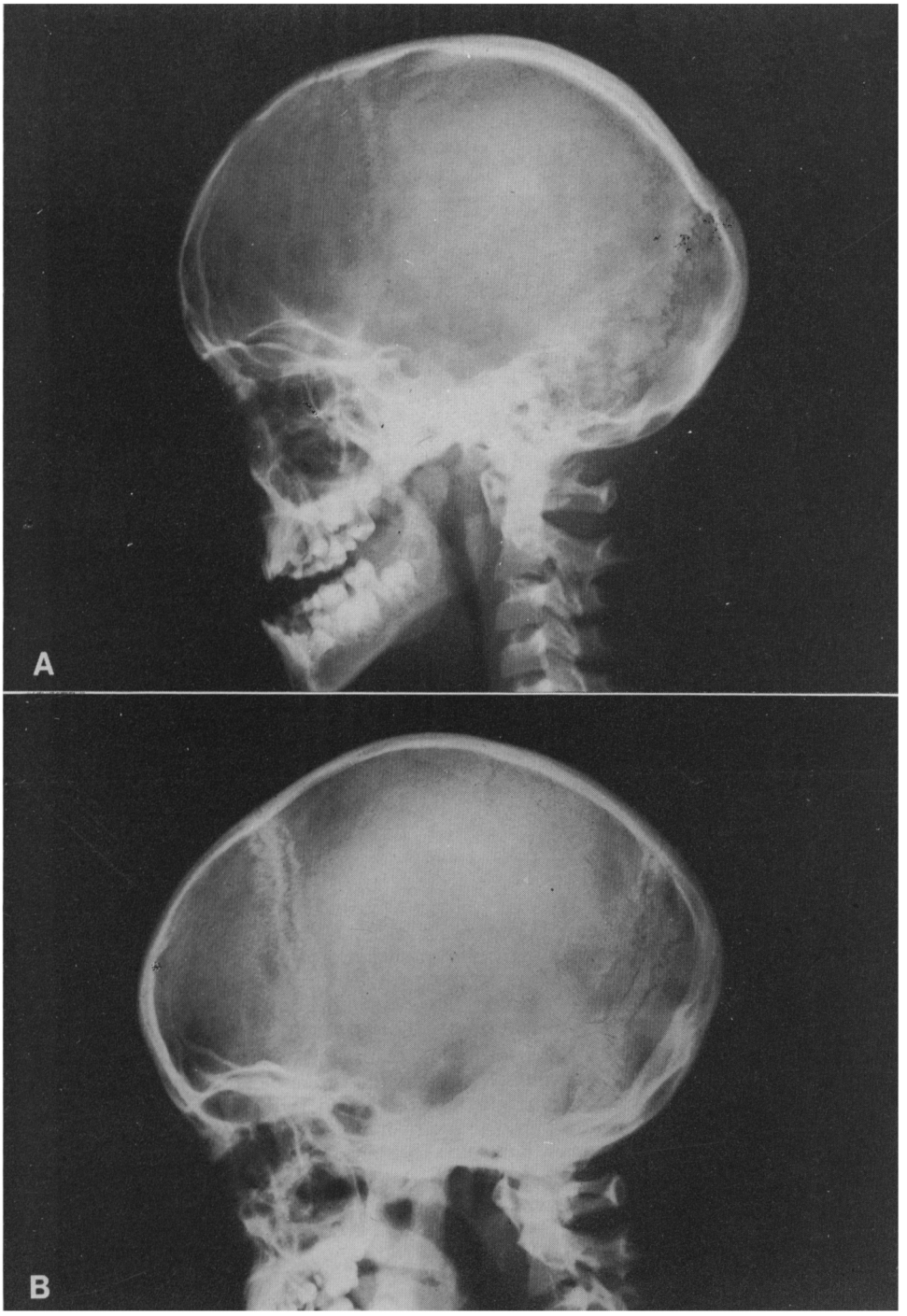

Fig. 2

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body, and her P. A. film shows a persistent frontal (metopic) suture and a relatively undifferentiated frontal sinus area. None of these features is present in the corresponding roentgenograms of Twin A, in whom the frontal sinuses are further developed. Since the skull films of the two girls seem to have been made at slightly different tube-target distances, the apparent dimensions of various structures visible in them are not directly comparable. The films do, however, show marked differences in the shape of their skulls and bony orbits.

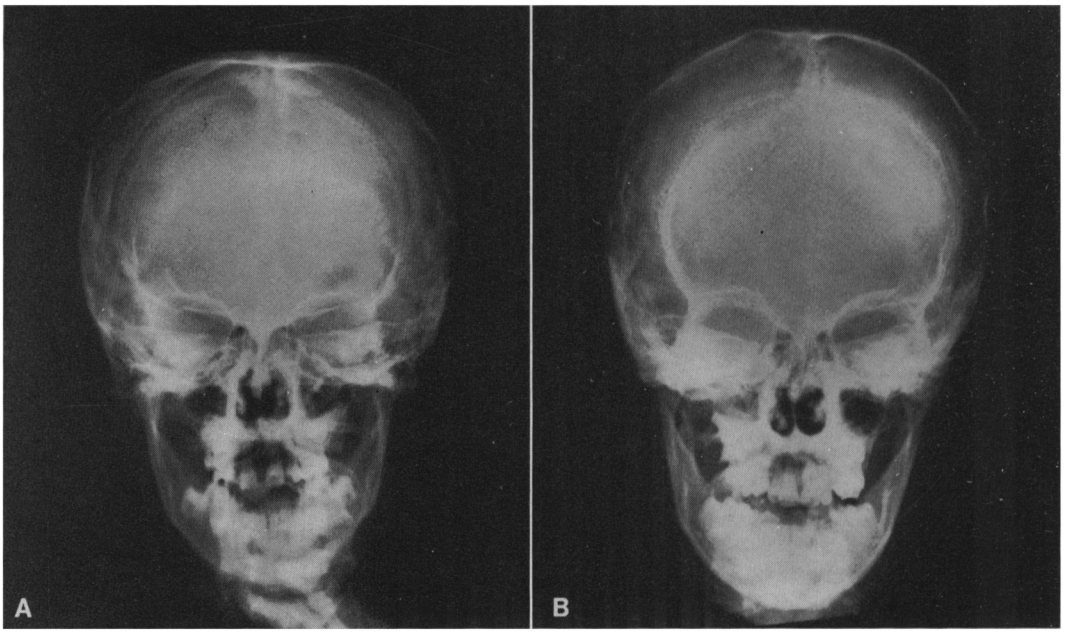

Fig. 3

As can be seen in the hand-roentgenograms shown in Fig. 4, Twin A had, at that time, markedly dysplastic middle phalanges and clinodactyly of both fifth fingers. Her more mongoloid sister did not have these digital defects. It will be noted, too, that both second metacarpals of Twin A (but not those of Twin B) had a basal as well as the normal distal epiphysis.

Dr. Charles S. Culotta, then of the Department of Pediatrics at Yale University School of Medicine, who examined the twins, reported that, though both children were definitely mongoloid, the various stigmata of mongolism were more numerous and more marked in Twin B than in Twin A. In addition, Twin B had a loud blowing systolic murmur. Both girls had fine blond hair and very similar grayish-blue eye color. Twin B, but not Twin A, had some Brushfield's spots in the iris of the left eye.

Dr. Bronislaw Symkowski, of Bridgeport, Connecticut, who had delivered the twins, kindly provided a copy of the abstract of the case taken from the records of the Bridgeport Hospital, where the mother had been confined. The pertinent parts of that abstract follow.

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BRIDGEPORT HOSPITAL
Bridgeport, Conn.

ABSTRACT

NAME: Mrs. M. V.
ADMITTED TO THIS HOSPITAL: 10-9-28. AGE: 41 years.

DIAGNOSIS: Parturition L.O.A. (Twins).
OPERATION: Low forceps.
DISCHARGED: 10-29-28. CONDITION ON DISCHARGE: Recovered.

HISTORY:

C.C. Parturition - para. 9.
P.I. No signs or symptoms of toxemia during pregnancy. Patient enjoyed fairly good health.
P.H. No serious illnesses.
4 normal deliveries.
2 miscarriages.
Last CTA January 25, 1928.

PHYSICAL EXAM:

HEART Normal
LUNGS Clear - no rales.

OBSTETRICAL CHART:

Number of child 9. Character of former labors normal. 2 miscarriages - unknown cause. First menses 12 1/2 years. Last menses January 25, 1928.

Admitted to labor room at 4:45 A.M., October 9, 1928. Bladder empty - bowels empty - enema at 12 hours. Temperature 98.6 - pulse 86 - respirations 20.

First stage began at 2 A.M. Position L.O.A. Pituitrin 1 cc given intramuscularly. Foetal heart 134 - left lower quadrant. Uterine contractions fairly good. Membranes ruptured spontaneously. No. 1 birth at 9:28 A.M. - No. 2 at 9:36 A.M. Ergot - drams 1 - hemorrhage slight. Female twins.

Delivery by very low forceps of a living female, No. 1, at 9:29 A.M. - position L.O.A. - delivered normally of a living female, No. 2, 9:36 A.M. Double footling presentation - double ovum twins. No lacerations or sutures. Placentae expressed spontaneously at 9:40 A.M. Ergot, dram 1 at 9:41 and 10:10 A.M. Pituitrin, 1 cc 7:56 A.M. and 9:06 A.M.

It will be noted that Dr. Symkowski recorded them as "double ovum" twins and stated that the "placent*ae* expressed spontaneously." It is not surprising that, when the case was discussed with him, more than ten years after the birth of the twins, he had no clear recollection of the character of the other membranes.

In an interview with the mother at her home, in 1939, some additional information about her reproductive history was obtained. According to her account at that time, the age, sex, and birth order of her children were as follows:

Sex:	F	M	M	M	F	F	M	F	FF
Age: (in years)	29	Miscarried at 7 months	27	25	21	20	18	13	10

She mentioned only one miscarriage, but the hospital Abstract lists two, which is probably correct. This discrepancy was not noted at the time of the interview, because the interviewer had not yet seen the Abstract.

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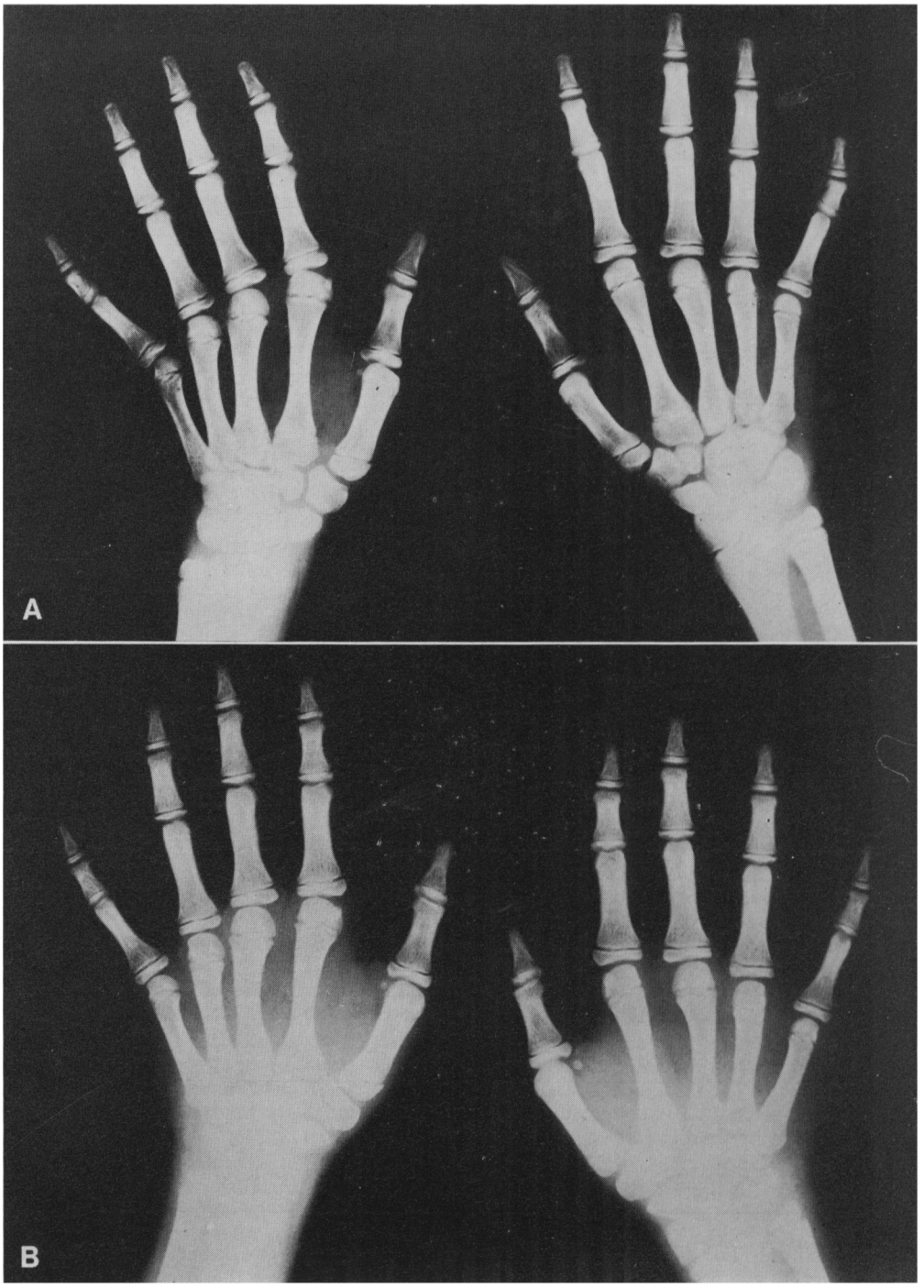

Fig. 4

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Because of the physical dissimilarities of the twins described above, I had considered them to be DZ and had described them as such in lectures to medical students and in other discussions of twinning and of mongolism in which there was occasion to refer to them. Several years ago, these photographs and hand-radiographs of the twins were among the materials used to illustrate a paper on the incidence of dysplastic middle phalanges of the fifth finger that was presented at a symposium held at the National Institutes of Health, in Bethesda, Maryland (Greulich 1969). The discussion evoked by the paper suggested the desirability of trying to obtain some additional information about the twins that might serve to disprove or to confirm my original view of their zygosity.

Locating this pair of twins after more than thirty years — if, indeed, they were still alive — seemed, at first, a rather dubious enterprise. Thanks, however, to great good fortune and to the help of Dr. Francis E. Martin, Medical Director of the Mansfield Training School, Mansfield Depot, Connecticut, where the twins had spent a number of years, it was learned that, some years earlier, they had been transferred to the Southbury Training School, Southbury, Connecticut. Dr. Herman Yannet, the Medical Director, and Dr. W. Roy Breg, Jr., Senior Physician of the Southbury institution, kindly reexamined the twins and obtained blood samples, photographs, finger and palm prints, hand and wrist roentgenograms, and karyotype analyses of them, in a further effort to determine their zygosity. The twins were 45 years old when those follow-up studies were completed.

THE NEW FINDING

The twins were reexamined by Dr. Yannet, on 26 September 1972, when they were 43 years and 11 months old. Pertinent parts of the letter in which he reported some of his findings follow.

"Both twins have the regular 21-Trisomy type of mongolism. They both now have the same eye color, which is medium gray. Twin B has a cataract on the right, which apparently, is not interfering significantly with her vision. Twin A has very dark brown hair. Twin B has a total alopecia. Unfortunately I don't remember the color of her hair before it disappeared, which was many years ago. The nose of both twins is definitely peculiar. Twin A's has a very striking bulbous tip. In Twin B this is much less striking and appears more as a downward turning of the tip of the nose. Unfortunately, none of the nurse or attendants has seen any of the twins' sisters or brothers recently enough to remember anything about their appearance." [This was in response to my question as to whether or not their unusual nose shape was shared by any of their siblings.]

The photographs reproduced in Figs. 5*a* and 5*b* were made when the twins were about 44 years old. Because of her alopecia, Twin B usually wears a wig, which somewhat exaggerates the resemblance between her and her sister. Later, she generously permitted us to photograph her without the wig, and her sister kindly rearranged her own hair so as to facilitate a comparison of their head and facial profiles (Fig. 5*c*). The same illustration shows, also, the severity of Twin B's alopecia and the associated fine texture of her hair as compared with that of Twin A. The latter feature can be seen even more clearly in Fig. 6, which is intended chiefly to show the differences in the ear form of the two girls.

The prominent frontal bosses of Twin B and the open metopic suture visible in the radiograph of her skull are probably attributable to the incomplete differentiation of her frontal bones.

The new roentgenograms of the hands and wrists (Fig. 7) are somewhat clearer than the original ones. This makes it easier to compare the size and shape of the individual bones of one twin with the corresponding bones of the other. The characters used in making this comparison are those which have been found useful in establishing individual identification from hand roentgenograms (Greulich 1960).

The differences in the form of the individual carpals, e.g., capitate, hamate, trapezium, and trapezoid, are much more marked than those which we found in normal twins who were considered to be MZ (Greulich 1960: pp. 763-764). This comparison was directed especially to the carpals, because their shape is probably less affected by the subject's mongolism than are the length and proportions of the metacarpals and the phalanges. The reduction in the length of the styloid process of the ulnae

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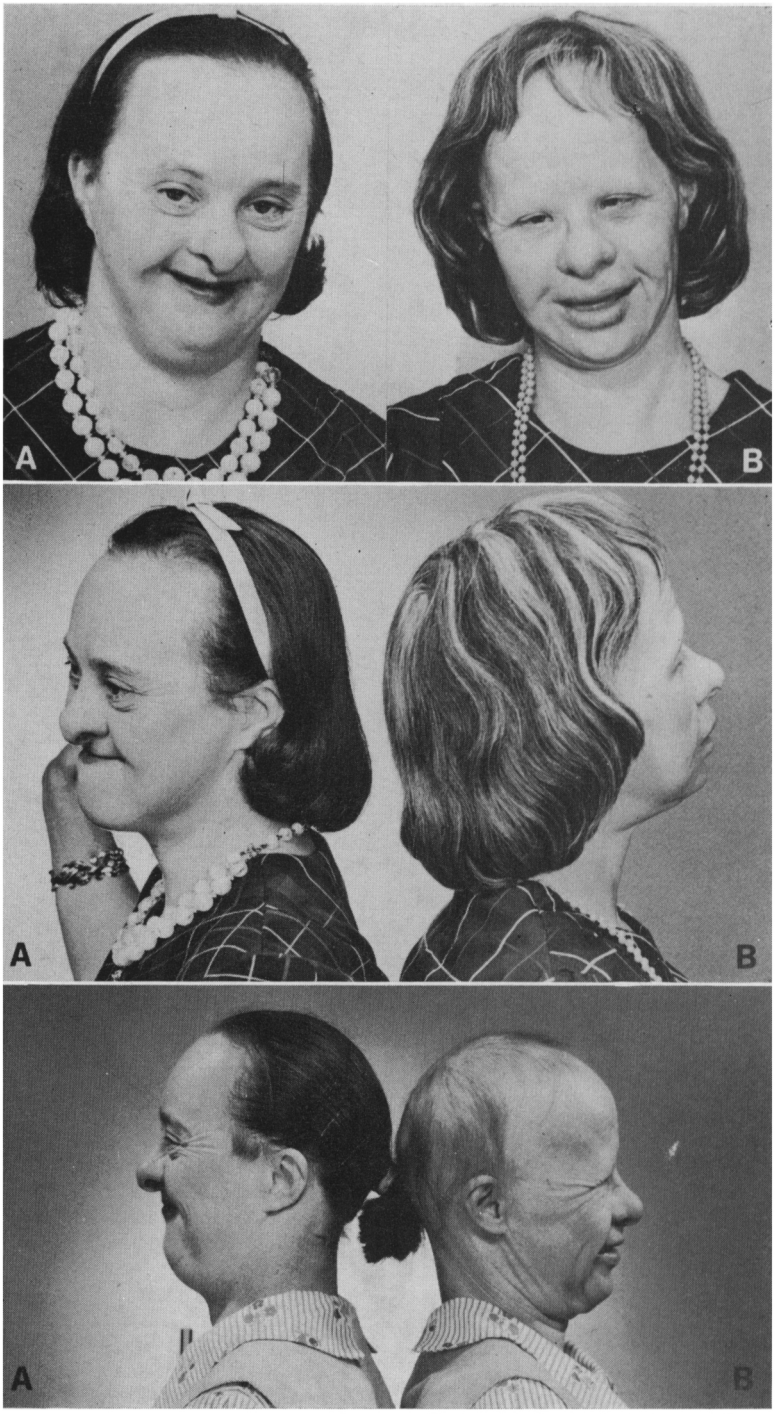

Fig. 5

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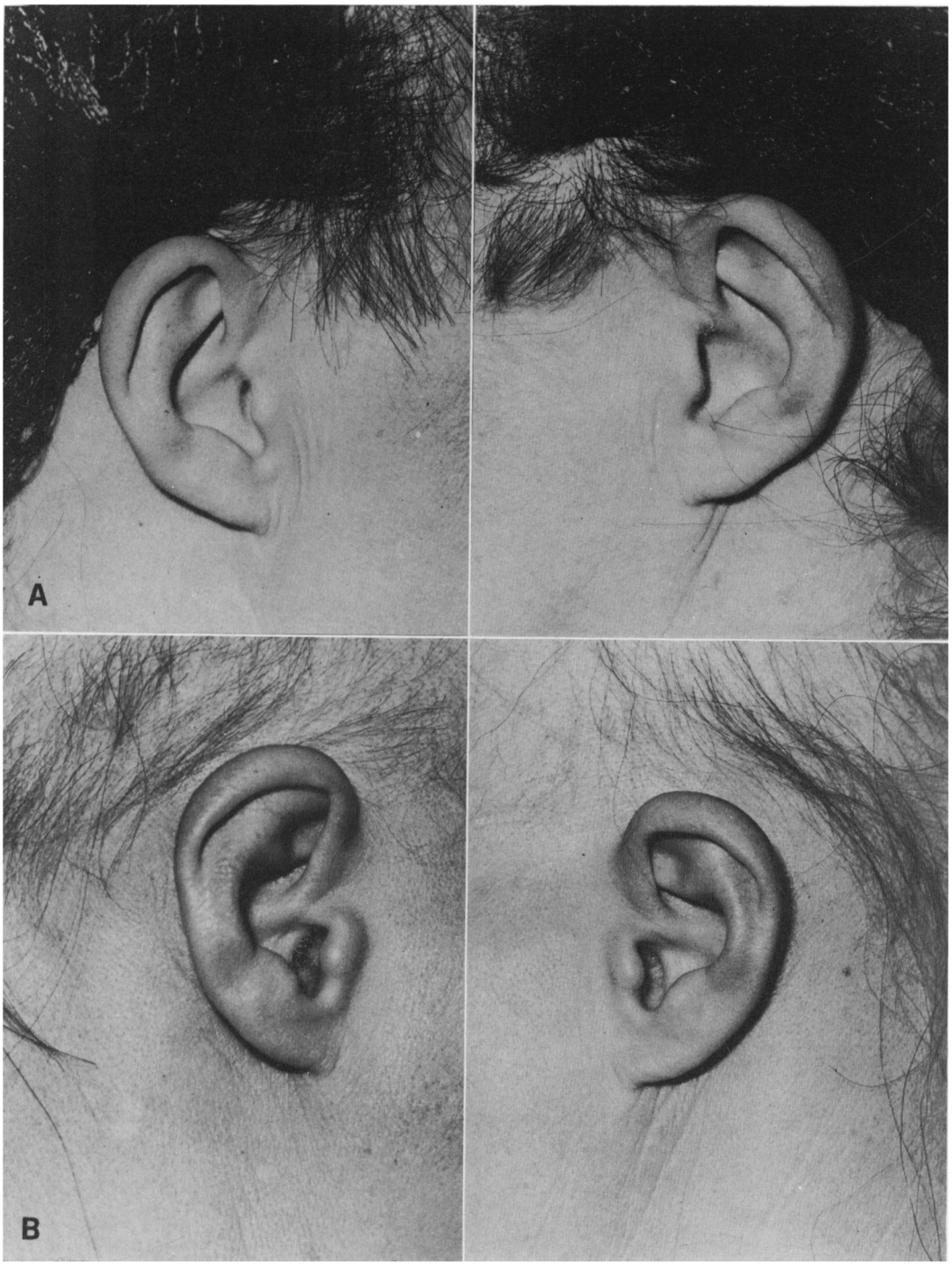

Fig. 6

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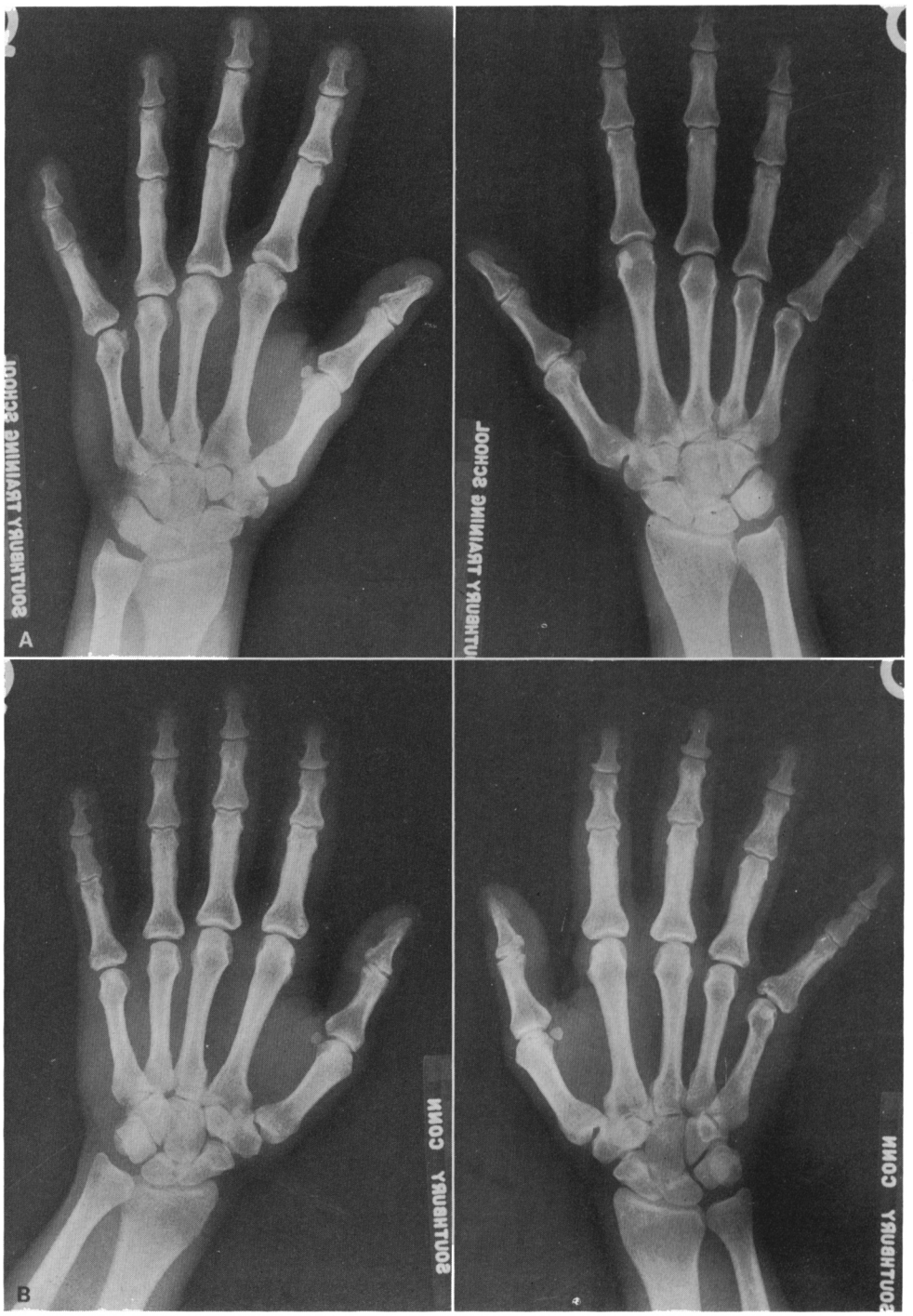

Fig. 7

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in Twin B, as compared with that of Twin A, is probably attributable to the more severe mongolism of the former. Our studies of the hand-films of other mongoloids suggest that this trait occurs more frequently and is more pronounced among the lower- than among the higher-grade Down's syndrome patients.

The blood studies and the interpretation of the dermatoglyphics of these twins were made by Chris C. Plato, then of the Children's Diagnostic and Study Branch of the National Institute of Child Health and Human Development, National Institutes of Health, Bethesda, Maryland. The following is his account of his findings.

Blood Groups. Blood specimens from the twins were collected and typed for most of the routine blood groups. The blood types of each twin are shown below.

Twin A: A_1, MMS$\bar{s}$, Mg^-, P_1^+, $\bar{c}\bar{c}$DEe, $\bar{k}\bar{k}$, $Kp^{(a-b+)}$, Lu^{a-}, Le^{a-}, Xg^{a-}

Twin B: A_1, MN$\bar{s}\bar{s}$, Mg^-, P_1^-, $\bar{c}\bar{c}$DEe, $\bar{k}\bar{k}$, $Kp^{(a-b+)}$, Lu^{a-}, Le^{a-}, Xg^{a-}

The differences observed in the MNS system as well as that in the P_1 indicate that they are of dizygotic origin.

Haptoglobins. The haptoglobin types of both twins were evaluated through starch gel electrophoresis and both were found to be of Hp 2-2 phenotype.

Dermatoglyphics. The dermatoglyphic evaluations included both finger- and palm-print comparisons. Unfortunately, despite repeated efforts, no satisfactory prints could be obtained for ridge counting. This was mainly due to the age of the subjects. The results are summarized in the Table. The main lines and other dermatoglyphic markers were graphically drawn and presented in Fig. 8.

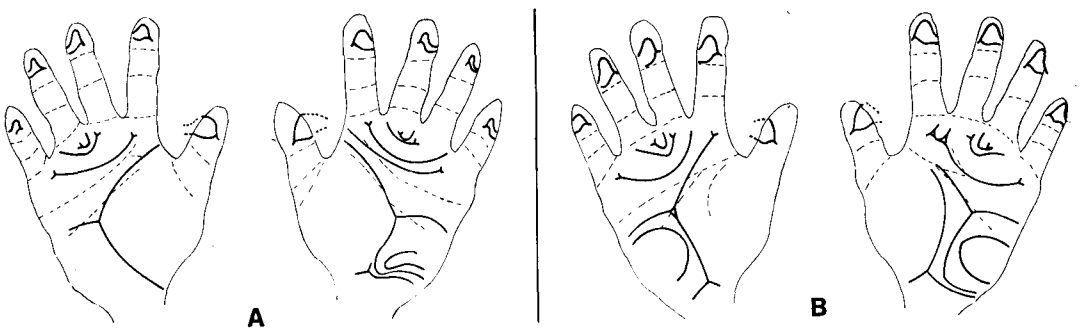

Fig. 8

Both twins show most of the cardinal dermatoglyphic signs of mongolism, i.e., excess of digital ulnar loops, wide atd angle due to the distal location of the triradius, and presence of pattern (loop) in the third interdigital area of the palm. Twin B furthermore shows bilateral ulnar loops in the hypothenar area which are accompanied with the presence of accessory axial triradii and bilateral simian creases (aberrant type in the right palm). Twin A does not have a simian crease in either palm, and only the right palm shows a hypothenar pattern.

The main line terminations are of interest because of their unusually transverse direction. This

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TABLE

DERMATOGLYPHIC ANALYSIS OF THE TWO MONGOLOID TWINS

	LEFT					RIGHT					Pattern Intensity Index
FINGER PRINT PATTERNS											
	I	II	III	IV	V	I	II	III	IV	V	
Twin A	U	U	U	U	U	U	U	U	U	U	10
Twin B	U	U	U	U	U	U	U	U	U	U	10
MAIN LINE TERMINATIONS											
		D	C	B	A		D	C	B	A	
Twin A		12	9	7	6		13	11	9	7	
Twin B		11	9	7	2/5		11/12	9	7	11/6	
PALMAR CONFIGURATIONS											
	Hypothenar	Thenar T	II	III	IV		Hypothenar	Thenar T	II	III	IV
Twin A	0	0	0	L	0		U	0	0	L	0
Twin B	U	0	0	L	0		U	0	L	L	0
MISCELLANEOUS PALMAR OBSERVATIONS											
	Simian line	Sydney line	Accessory triradius	atd angle			Simian line	Sydney line	Accessory triradius	atd angle	
Twin A	—	—	—	66°			—	—	t	37°/63°	
Twin B	Complete	—	t	32°/88°			Aberrant	—	t, II	43°/81°	

is especially true in Twin A. It should also be noted that the loop in the III interdigital area of the right hand of Twin A is a result of the ulnar direction of line C instead of the customary radial.

In summarizing, the dermatoglyphic differences agree with the blood group findings that these twins are of dizygotic origin.

In 1970, when Mr. Plato first studied the twins' blood, he also arranged to have a portion of the same samples typed by another investigator, at a different laboratory. The latter's findings agreed with his own.

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Because the validity of the differences disclosed by the 1970 blood studies of the twins seemed crucial in determining their zygosity, it was mutually decided to repeat the tests before this report was submitted for publication. Accordingly, Dr. Breg collected blood samples from the twins again, in 1974, and sent them on to Mr. Plato, who with Mr. Wilson Leyton, of the National Institute of Dental Research, Bethesda, Maryland, independently typed the new samples. Their findings agreed with each other's and with those made four years earlier.

Since three different investigators, using test sera from five different sources, obtained the same results in 1970 and in 1974, it seems reasonable to conclude that the reported serological differences between the twins are valid ones.

DISCUSSION AND CONCLUSION

Determining the zygosity of like-sexed twins on the basis of similarities or differences in the various external physical features usually employed for that purpose is especially difficult in subjects with Down's syndrome, because the final form of those characters can be, and frequently is, modified or even distorted in the presence of mongolism.

The dermatoglyphic pattern of a person with Down's syndrome, for example, is certainly not the same as it would have been in the absence of his mongolism: indeed, the dermatoglyphic pattern is modified in such a characteristic way in mongolism that its resulting form has become an important feature of the syndrome.

The extent to which the normal dermatoglyphic pattern is thus modified in a pair of like-sexed, mongoloid twins, reduces correspondingly its usefulness as an aid in determining their zygosity. This is equally true of any other genetically determined trait or character, the expression of which can be appreciably modified or distorted in mongolism. And such distortion can be rather widespread in persons with this syndrome; for it appears that the mongoloid is derived from a zygote which lacks either the capacity to generate, or to respond properly to, those stimuli which normally implement and direct embryonic, fetal, as well as postnatal development (Greulich 1973).

Many internal characters also are comparably modified in mongolism and become quite as valid features of the syndrome as the external ones. The most constant of these internal stigmata of mongolism are, of course, those brain defects which manifest themselves in the mental deficiency that is present to some degree in all mongoloids.

Some less constant internal defects associated with Down's syndrome are congenital cardiac and other vascular anomalies; the immunological inadequacies responsible for the mongoloid's reported increased susceptibility or low resistance to various infections, especially to those of the respiratory tract (Penrose and Smith 1966); the persisting relative immaturity of some white blood cells as suggested by the low incidence of multilobular nuclei among their neutrophilic leukocytes (Mittwoch 1958); and a comparable immaturity of their erythrocytes that expresses itself in macrocytosis and increased cell volume (Naiman et al. 1965). Perhaps, one might add to this list the heightened susceptibility to leukemia as indicated by an increased incidence of that disease among mongoloids, especially during early childhood (Krivit and Good 1957, Stewart 1961).

In view of the rather widespread involvement of the blood vascular system in Down's syndrome, it is remarkable that the various blood factors that have been found so useful in determining the zygosity of normal like-sexed twins should entirely escape the distorting influences of the mongoloid state — if, indeed, they do. Or can the presence of Down's syndrome occasionally prevent the full and proper expression of the genetic determiners

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for some blood factors, in much the same way as it inhibits or distorts the normal development of some other genetically determined characters to produce the mongoloid phenotype?

The determination of zygosity in a pair of like-sexed mongoloid twins can be only tentative, and not conclusive, for it has to be based on an evaluation — at least partly subjective — of similarities and differences between them in genetically determined traits whose expression has been variously modified by their common mongolism.

In their totality, the differences noted in this pair of twins seem to support the conclusion that they are dizygotic. I found no equally persuasive evidence of their monozygosity. The observed differences in some of their blood factors, in their dermatoglyphic patterns, in certain of their skeletal features, in the severity of their mongolism and other differences described in this paper, argue strongly against that assumption.

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RIASSUNTO

Sindrome di Down Concordante in due Gemelle DZ Studiate all'Età di 10 e 43 Anni

Una coppia di gemelle mongoloidi è stata studiata all'età di 10 anni e nuovamente circa 30 anni dopo. Sono state attentamente raffrontate somiglianze e diversità nelle caratteristiche utili nella determinazione dello zigotismo.

Benché alcune di queste caratteristiche possano essere, e frequentemente siano, notevolmente modificate in presenza di mongolismo, ed in tal modo perdere parte del loro valore, le differenze notate in queste gemelle, sia da bambine che da adulte di mezza età, sembrano consentire la conclusione che si tratti in realtà di gemelle dizigotiche.

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RÉSUMÉ

Syndrome de Down Concordant chez Deux Jumelles DZ Examinées à l'Age de 10 et 43 Ans

Un couple de jumelles mongoloïdes a été examiné à l'âge de 10 ans et de nouveau environ 30 ans plus tard. Les ressemblances et diversités dans les caractéristiques utiles à la détermination de la zygosité ont été soigneusement confrontées.
Bien que certaines de ces caractéristiques puissent être — et elles le sont fréquemment — remarquablement modifiées en présence de mongolisme, et de cette façon perdre une partie de leur valeur, les différences observées chez ces jumelles, examinées enfants puis adultes d'âge moyen, semblent permettre de conclure qu'il s'agit bien de jumelles dizygotiques.

ZUSAMMENFASSUNG

Konkordantes Downsches Syndrom bei ZZ Zwillingsschwestern, die mit 10 und 43 Jahren untersucht wurden

Bei zwei mongoloiden Zwillingsschwestern wurden erstmalig im Alter von 10 J. und sodann ungefähr 30 J. später die Aehnlichkeiten und Abweichungen der für die Eiigkeitsbestimmung nützlichen Merkmale untersucht.
Obwohl einige dieser Merkmale bei Mongolismus erheblich verändert sein können und es oft auch sind, wodurch sie einen Teil ihres Aussagewertes einbüssen, so schienen doch die bei diesen Zwillingsschwestern sowohl im Kindes- als im Erwachsenenalter beobachteten Unterschiede die Feststellung der Zweieiigkeit zu gestatten.

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