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WILLIAMS SYNDROME AS A MODEL OF GENETICALLY DETERMINED RIGHT-HEMISPHERE DOMINANCE

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Studies were carried out on the dermatoglyphics (skin ridge marks) on the hands of children with Williams syndrome; this is an inherited disease with cardiovascular pathology and a characteristic facial phenotype ("elf" facies), along with specific mental and cognitive disturbances. The results suggest a characteristic dermatoglyphic type with the presence of complex whorls on the fingers and a clear predominance of marks of greater complexity on the left hand; this is a very rare trait in normal people and in those with other inherited nervous system disorders. The features of the dermatoglyphic pattern serve as a characteristic marker of a genetically determined state of the human central nervous system, and suggests directions for neurophysiological studies of children with Williams syndrome as a unique model for analysis of higher nervous function in humans.

Key words: Williams syndrome, dermatoglyphics, interhemisphere brain asymmetry.

The modeling of a variety of processes in humans and animals represents a cardinal direction in experimental physiology. The model may be the organism itself or, better, some kind of defined, constitutional type of organism in which studies of specific responses to defined experimental treatments and of adaptation to particular environments provides another approach to understanding the mechanisms regulating homeostasis. Probably the easiest types of humans to diagnose are the types described in the terms of syndromology, which is a relatively young area of contemporary clinical medicine, with great potential, in which studies involve numerous inherited and genetically determined properties of development (which are sometimes so profound as to be developmental defects). Williams syndrome is one such type; this is a relatively common inherited state (the American Williams Syndrome Association included more than 1000 patients in 1990 [7]), and produces an unusual facial phenotype ("elf" facies) and a number of defects in the cardiovascular system, along with specific mental and cognitive abnormalities presenting as a typical continuum ranging from pronounced mental retardation to a nearly-normal level of intellect. The features of the mental state of patients with Williams syndrome are specific and can even be regarded as a unique complex of traits ([9] and others), and include: (1) disturbance of sensory integration with hypersensitivity to sound and "gravitational anxiety" [6]; (2) hyperreactivity with emotional lability, impulsiveness, attention deficit, intrusive communicativeness; (3) elevated anxiety and fear of novelty; (4) disturbances in expressive and impressive speech; (5) learning difficulty, especially in mathematics, but with relative ease in learning to read; (6) a good musical ear and sensitivity to rhythm [4]. These features suggest a particular neurophysiological type, whose morphophysiological basis is a particular central nervous system structural type. Dermatoglyphics may make a contribution to the study of this condition. The importance of the dermatoglyphic approach is that firstly, the ridged skin arises from the same embryonic rudiments as central nervous system structures [11]; secondly, dermatoglyphic signs do not change through life and are relatively easy to classify for a given individual [3, 11]. However, it should be emphasized that in our view, the complete dermatoglyphic type of an individual should be assessed, a process which is not followed in the formal analysis of particular skin ridge signs used in traditional dermatoglyphics, a limitation which may explain the lack of progress in this area of knowledge. Such an approach provides a good indication of the human as an entire system in terms of his "individual and historical development" (Schmalhausen).

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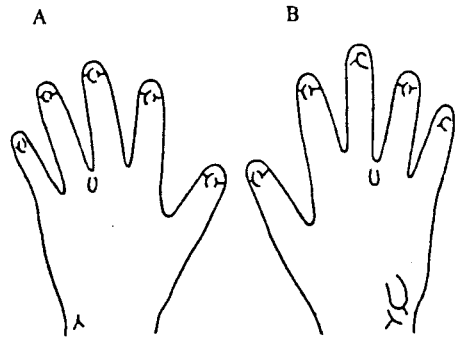

Fig. 1. Fingerprint pattern in Williams syndrome. *A*) Left hand; *B*) right hand.

METHODS

Dermatoglyphic features were studied in 16 children with Williams syndrome (six boys and ten girls), aged 3 to 10 years. The control group consisted of 176 children (92 boys and 84 girls) at Moscow schools, aged 5-12 years. Prints were taken from the fingers and palms using printer's ink onto paper and were analyzed at the fingertips and palmar fields using the classical method of Cummings and Midlow [11].

RESULTS AND DISCUSSION

These studies showed that the dermatoglyphic pattern of the hands of children with Williams syndrome consisted of a typical dermatoglyphic type (Fig. 1, Table 1), characterized by a predominance of correctly formed, symmetrical, complex (with two triradii) ridges on the fingers, consisting of whorls with high ridge counts. Marks of greater complexity were located predominantly on the left hand. Interdigital asymmetry was most frequently seen on the third and/or fifth fingers. Two children had the so-called "monomorphic hands," in which the skin ridge patterns were identical on all fingers (one case with whorls, the other with ulnar loops); however, the ridge count in these cases was greater on the left. Markings in the palmar fields were very insignificant, and consisted of axial triradii and bent palmar creases which were normal.

The predominance of whorls is a relatively rare trait in normal people (Table 1), and the predominance of markings of greater complexity on the left hand is even rarer (Table 1). These features and the absence of abnormalities in the dermatoglyphic pattern suggest that the dermatoglyphic type of children with Williams syndrome is a rare variant of the normal pattern. As already noted, fingerprints can be used as a marker of the central nervous system type. This suggestion is supported by their common embryonic origin and also by the relationship between the complexity of fingerprints and the structure of the nerve endings in the terminal phalanges of the fingers, as shown by the classical studies of Bonnevie [8], and whose central determination is not in doubt. On the other hand, extensive clinical experience [11] supports the existence of a relationship between disturbances in central nervous system development and alterations in the characteristics of dermatoglyphic marks. Additionally, healthy children showed a correlation between particular dermatoglyphic patterns and resting EEG traces, in that the more complex fingerprints were associated with higher power levels in the high-frequency EEG range [1]; this may reflect the genetically determined features of brain structure in these children.

The fact that more complex dermatoglyphic marks are more frequent on the right hand, which is dominant in most people, has been noted in numerous studies [3, 5]. In turn, the dominant role of the left hemisphere in higher (primarily speech) brain functions is well known. These observations provide the basis for the concept of hemisphere dominance. Current data on morphological asymmetry (for example, the greater area of the temporal plane entering Wernicke's area on the left [10, 12]) also reflect the dominance of the left hemisphere in brain activity. Dermatoglyphics in Williams syndrome provides us with a mirror image of the usual pattern of predominance of skin ridge complexity. This leads to the question of whether this dermatoglyphic pattern, especially the asymmetry in fingerprints, is an indicator of a unique central nervous system type in

TABLE 1. Distribution of Fingerprint Features (dermatoglyphics) in the Hands of Children with Williams Syndrome and Healthy Children

Dermatoglyphic feature	Williams syndrome		Control group	
	$n = 16$	%	$n = 176$	%
8-10 whorls	9	54*	26	15
5-7 whorls	3	19	27	15
9-10 loops	3	19	39	22
5 arches	1	6	4	2
Asymmetry $D_s > D_d$, third finger	7	44*	12	7
Asymmetry $D_s > D_d$, fifth finger	6	38*	10	6
Asymmetry $D_s > D_d$, any finger	12	75*	15	9

Notes. D_s is the number of deltas (finger triradii) on the fingers of the left hand, D_d is the number of deltas (finger triradii) on the fingers of the right hand. *) $p < 0.001$ (χ^2 criterion).

children with this syndrome. This would be a type in which a significant characteristic is a more complex morphological organization of the right rather than the left (as in most people) hemisphere. This suggestion clearly needs careful verification and better evidence, and indicates the need for a re-examination of the nature of the mental state of these children, especially in terms of the level of intellectual development. The absence of abnormalities in the dermatoglyphic pattern in Williams syndrome suggests that there is no deep abnormality in brain structure in these children, in whom the diagnostic significance of pathological fingerprint features is well known [11]. This suggestion is also supported by the fact that children with Williams syndrome, despite their learning difficulty, can nonetheless generally adapt to normal schools [4]. At the same time, it would be no great exaggeration to say that current tests for intellectual level generally assess the capacity of the left hemisphere. It is possible that many behavioral features, including some which may not yet have been described, of Williams syndrome children will reflect this unique interhemisphere asymmetry. It should be borne in mind that a significant proportion of our knowledge of the functions of the right hemisphere has been obtained from pathological conditions (infarcts, brain insult, tumors, traumatic lesions, etc.) [2]. Williams syndrome may thus provide a model for starting a new stage in the physiological study of this terra incognita, the right hemisphere.

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