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Un caso de polidactilia en un embrión de 22 mm.

El caso presentado en este trabajo, si el autor está bien informado, es el más joven mencionado hasta el presente, debiendo su significación principalmente a este hecho. Además suministra datos adicionales interesantes sobre el problema del primer hueso metacarpiano y sobre el problema más general del polidactilismo. En este caso el autor encuentra, sin embargo, pocas pruebas concluyentes en favor de las teorías relacionadas con ambos problemas. El único punto valioso que el autor puede ofrecer es que el presente caso lleva el problema del polidactilismo desde el catálogo de los factores causativos extrínsecos al dominio de los factores formativos tempranos.

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A CASE OF UNILATERAL POLYDACTYLY IN A 22-MM. EMBRYO

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ONE FIGURE

In the laboratory of the author there is an embryo 22 mm. in length, with an extra digit on the right hand at the base of the metacarpus of the thumb. It was noted by Doctor Streeter while making measurements of the embryo, and he marked it as being, from all available records, the youngest case thus far discovered. This fact seems to warrant the author in making a study and report of it, with a brief survey of the literature on the subject.

The embryo came into the laboratory without data as to its parents or details of its appearance. It has a crown-rump length of 22 mm. and is well formed in every detail, with membranes in normal state. The two hands are similar in every respect, except that the thumb on the right hand is a trifle smaller than the left thumb. The breadth of each hand, midway across the palm, is 2 mm., and the length, from a line across the wrist just back of the thenar eminence to the tip of the middle finger, is 2.5 mm. The right thumb is 1.1 mm. long and 1 mm. thick. Attached to the base of its metacarpus is the extra digit. It stands out from the surface at right angles and is uniform in size and shape. Near the tip it bends slightly forward and terminates in a blunt rounded point. It is 1 mm. long and 0.6 mm. thick. Serial cross-sections were made of the whole forearm and hand beginning at the tip of the fingers and going to the elbow. The sections were cut 10μ thick after staining the specimen in toto in cochineal.

Fig. 1 Outline drawing from reconstructed model. $\times 22$. A. Surface view showing position of extra digit. B. Cartilaginous skeleton showing relationship of extra digit to other skeletal units

Microscopic study shows the cartilages of the wrist, hand, and fingers present. All the separate units of the parts are in place and are still in process of cartilage formation. Each piece shows a peripheral zone of condensed cells beginning to show the features of a perichondrium, and a central core characterized by a network of hyaline strands, in the meshes of which may be seen large cartilage cells. The skeleton of the extra digit is a single piece of cartilage having the above characteristics. It is a separate unit with close relationship to the base of the metacarpus, and not an offshoot or spur from it.

Two muscles are contained in the digit, a volar and a dorsal muscle. The one on the dorsal side extends through the mesenchyma out toward the tip, but ending short of it. Its origin and identity are uncertain; probably it comes from the outer head of the first dorsal interosseous muscle. The volar muscle can be more certainly identified. It arises from the abductor pollicis brevis and extends to near the tip of the digit on the palmar side. Nerves cannot be identified and vessels are seen only as undifferentiated capillaries.

In going through the literature, no cases are cited of polydactyly in the young embryo; all are late foetuses or postnatal cases and the majority are in adult life. Salzer ('98), reporting two cases of triple phalangeal thumbs, opened the question as to which phalanx is missing in the ordinary thumb, the end phalanx or the middle phalanx. He quoted Windle, who made a general summary of such cases by expressing the opinion, with some reservations, that the extra phalanx must have occurred from the fact that the distal bone center of the first metacarpus developed independently. He referred to Uffelman's work, in which it is shown that at the capitellum of the first metacarpus a special bone center is present without ever existing independently. He thinks that this extra center may give rise to a supernumerary phalanx, which would be the main phalanx. Salzer quoted Pfitzner also, who seemed to hold that a triple phalangeal formation is, for a brief period, a normal state and that the double phalangeal thumbs and toes have come about in this manner, from the merging of the middle and end phalanges

gradually a typical and enlarged phalanx has resulted. In support of this he cites a case of a girl with a strikingly large end phalanx consisting of two pieces which have not been completely fused. Jurcie ('06) reported a case of hyperphalangea in both thumbs. In this report he took up the problem of whether the thumb should be considered as possessing normally three phalanges and no metacarpus, or a true metacarpus and only two phalanges. He cites the fact that the first bone of the thumb develops in the same manner as the phalanges, the head and shaft from a common primary center, and the base from an epiphysis. The development of the metacarpals is just the reverse, the base and shaft arise from a common primary center and the head from an epiphysis. He concludes, however, that from the manner of muscle attachment to the first metacarpus and noting the presence of the extra ossification center in some mammals, and occasionally in man, this is a true metacarpus and not a basal phalanx.

Stieve described a case of bilateral hyperphalangea ('15-16) and quoted Krause ('09), who objected to Pfitzner's work on the ground that it dealt with malformations, which was poor material for drawing such conclusions. In Stieve's case the end phalanx, which should be shorter than in normal diphalangeal thumbs, is especially long, and in Salzer's cases the two end phalanges show no proportionate shortening. He finds also that whereas there was restricted movement between the end and middle phalanges of Hilgenreiner's case, the movement in his case was especially free. Backed by these points, he comes to the conclusion, differing with his predecessors, that the end phalanx in the diphalangeal thumb is not a fusion of the middle and end phalanges. He seems to hold to this opinion more firmly when he notes that we are not dealing with perfect triphalangeal thumbs in reported cases, but the extra piece is only the radial side of the middle phalanx.

Hilgenreiner apparently has the most complete list of cases of hyperphalangeal thumbs and the most valuable data concerning the subject. According to him, 107 three-jointed thumbs, fifty-eight cases, had been noted to date ('10). He goes to

some length in discussing the condition and makes a definite classification of the cases. In the first class he would include cases where there is only partial separation of the end and middle phalanges—incomplete hyperphalangea. In class 2 he would include only such cases as show complete formation of the middle phalanx having a diaphysis and an epiphysis—true hyperphalangea. In these cases the thumb characteristics have been lost and the structure is more like an extra index-finger. This change of characteristics is proportionate to the degree of separation of the phalanges. He is quite firm in his opinion that the condition of diphalangeal thumb has arisen from a fusion of the end and middle phalanges in early stages of formation. He sees two possible explanations in these cases of anomalous thumbs, but does not pursue either at length. He speaks of a palingenesis of the middle phalanx traceable to some endogenous cause. This factor is mentioned by others also, Minoura and Prentiss. He also states that the supernumerary phalanx is not always to be explained as a palingenesis of the middle phalanx, since it may arise from the epiphysis of the second phalanx. In such cases triphalangeal thumbs appear only postpartem in the first, second, or third year of child life.

Prentiss ('03) has contributed to the subject a valuable piece of work, dealing with it in a somewhat broader fashion than Hilgenreiner, although his study of cases in man, especially of thumb variations, is rather too limited for valuable application here. He states that "variations are found chiefly in digits which are modified, rudimentary or vestigial." As to the causes of polydactyly he has this to offer: "in man it is not a definite number of extra digits but a tendency to duplication which is inherited" and "duplication of functional digits is probably caused by germinal variations alone."

After reviewing the above literature, which is, although brief, a general and inclusive statement of opinions on the subject, several problems present themselves or at least several aspects of the same problem. There appears to be a difference between hyperphalangea and polydactyly. The former is characterized by the presence of an extra phalanx, with the main disfiguration

of the thumb an increase in its length or a deflection to the ulnar side. The latter is marked by the presence of an extra digit, either large or small, fully formed or rudimentary, which stands apart from the other digits. The problem of di- and triphalangeal thumbs, as to how the normal thumb should be regarded and the origin of the extra phalanx in triphalangea, comes under the head of hyperphalangea. Here would be placed the cases of Jurcié, Salzer, Stieve, and some of Hilgenreiners. Under polydactyly would be included such cases as Prentiss', others of Hilgenreiner's, and the author's.

Another point is that variations in the thumb should be considered in a special class apart from variations in the other fingers, since here is found, in the normal state, a condition unlike that in the others.

Prentiss's statement that, "variations occur chiefly in digits that are modified," would lend emphasis to this, and Hilgenreiner seems to have considered it worth while to tabulate over one hundred cases of thumb variations.

Standing out prominently among these points is the problem of the causative factor in polydactyly and hyperphalangea, and how this factor operates. Upon this point the author's case presents some features, not noted in others, which might be considered a little more at length. Here is a clear case of polydactyly having an extra digit with a separate piece of cartilage. All the other units of the wrist and hand are present but still in the cartilaginous state. It is conclusive that the causative factor of splitting of a digit by an amnionic band is not operative here. Nor is the factor of an extra phalanx arising from a diverted center of ossification to be applied, since the digit is present before ossification appears anywhere in the hand. The specimen permits of an investigation further back than the beginning of ossification. Here might be applied the theory of palingenesis as suggested by Minoura, Hilgenreiner, and Prentiss, although the last author might not include it here. Or again, Prentiss' reversion to a not remote ancestral form, an inheritance, working through Mendel's law, might be applied. He states that "all cases of polydactyly, not due to external

causes, may be the result of inheritance." These two theories presuppose a parental state from which they may work, and the natural question arises as to what caused the first deviation in the original parent. 'Germinal variation' is an expression commonly used by investigators and especially by Prentiss when he says, "but the duplication of functional digits are probably caused by germinal variation alone." He further states that, "as to the cause of these germinal variations . . . we know little or nothing." In his comprehensive work, "Science and philosophy of the organism," Driesch put the essence of the matter in a more concrete form when he gave expression to 'feeling for form.' Just how far these expressions are usable is a question; they at least emphasize the fact that search must be made in the field of cytology and cytochemistry in dealing with the state of abnormal thumbs.

CONCLUSIONS

The author feels justified in drawing the following conclusions:

1. Hyperphalangea and polydactyly are sufficiently different to be put into separate classes.
2. The thumb offers sufficient significance to warrant separate study in its variations.
3. The author's case offers proof that some cases of polydactyly owe their origin to earlier causes than external factors or deviation of ossification centers.

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