

# HYPERPHALANGISM ACCOMPANIED BY SUPERNUMERARY EYPIPHYSES AND MUSCULAR DEFICIENCIES

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TWELVE FIGURES

This interesting developmental anomaly, was observed in the practise of Dr. Robert G. Reynolds of Palo Alto in March of 1916. The x-ray photographs accompanying this article were kindly taken by Dr. Reynolds, through whose courtesy I am enabled to report the case. As it usually happens in these instances the family history reveals related abnormalities. On the father's side of the family there have been several cases of abnormal development of the skeleton of the upper extremities. The paternal grandfather is said to have had syndactyly of the index finger and the thumb of the right hand, up to the interphalangeal joint of the thumb. The terminal phalanx of the thumb is said to have been flexed at right angles on the proximal phalanx so as to lie in the palm. The left hand and both feet were said to have been normal.

The father's oldest sister who died at the age of six is said to have had a deformity of the right hand similar to that of her father. Otherwise she was normal.

The father's second sister who died soon after birth is said to have had congenital absence of radius and ulna on the right arm and the father's older brother is said to have had similar deformities as the present case.

The father's hands both show muscular defects. On the left hand the thenar eminence is absent, and the opponens and abductor pollicis brevis apparently are lacking. The flexor brevis may be present. There are two groups of interphalangeal

and one of metacarpophalangeal sulci, in the thumb. Yet the x-ray photograph (fig. 1) shows that there are only two phalanges. The left thumb is straight and tapering; only about two-thirds normal size and the skin is smooth, shiny and atrophic. It is 6.4 cm. long and is rotated out a little. The mobility in the



Fig. 1 Left hand of father

second terminal phalanx of the thumb is somewhat limited. The width of the left hand in the line of the metacarpophalangeal articulations is 8.6 cm. There is no webbing of the fingers; the rest of the fingers of the left hand being apparently normal.

The thumb of the right hand is somewhat longer than that of the left, but the thenar eminence is also absent; the region being

marked by a decided flattening. The right thumb is 7.4 cm. long and is rotated in rather than out, as is the case with the left. It is tapering and atrophic as in the other hand, possessing a small but apparently normal nail. The pulp of the thumb is compressed from side to side and looks atrophied. There are two prominent sulci opposite the interphalangeal joint of the thumb and the width of the hand at the metacarpalphalangeal articulations is decidedly greater than in the left hand. The other fingers are normal. The width of the right hand in the line of the metacarpalphalangeal articulations is 9.2 cm. or 0.6 cm. more than the left. The medial surfaces of both thumbs, a little distal to the metacarpophalangeal articulations are marked by an oval callosity about 2 cm. in diameter and 0.5 cm. thick which forms a prominent pad. As shown in (fig. 2) the medial surface of the thumb is concave, the distal portion being bent medially, that is toward the ulna, the distal phalanx being directed medially at an angle of about 10 degrees.

In the x-ray photographs (figs. 1 and 2) there is a pronounced mortising of the basal ends of some of the metacarpals of the thumb with the basal ends of the metacarpals of the index fingers. This condition seems to be the result of a general narrowing of the wrist bones. In each hand the greater multangular is placed medially until its medial border is almost even with the medial border of the lesser multangular. Hence the position of these bones and the consequent position of the respective metacarpals may possibly account to some extent for the lack of mobility of the metacarpals of the thumbs. The proximal phalanx in each thumb is markedly concave on its palmar surface and has a slightly bent shaft. This is more noticeable in figure 2. These peculiarities in the thumb may be explained in part by the fact that the lack of mobility of the metacarpals has made it necessary to rotate the proximal phalanx medially in order to use the thumb in conjunction with the other fingers. As a result of this rotation we see the phalanges from a lateral view rather than a dorsal and therefore the concave surface is the ventral and not the medial surface of the phalanx. There doesn't seem to be anything to account for the three distinct

bony tubercles shown in outline in the x-ray plate on the ventral surface of the distal phalanx of each thumb (figs. 1 and 2).

The distal ends of the phalanges seem large and irregular. This is more noticeable in the right (fig. 2) than in the left

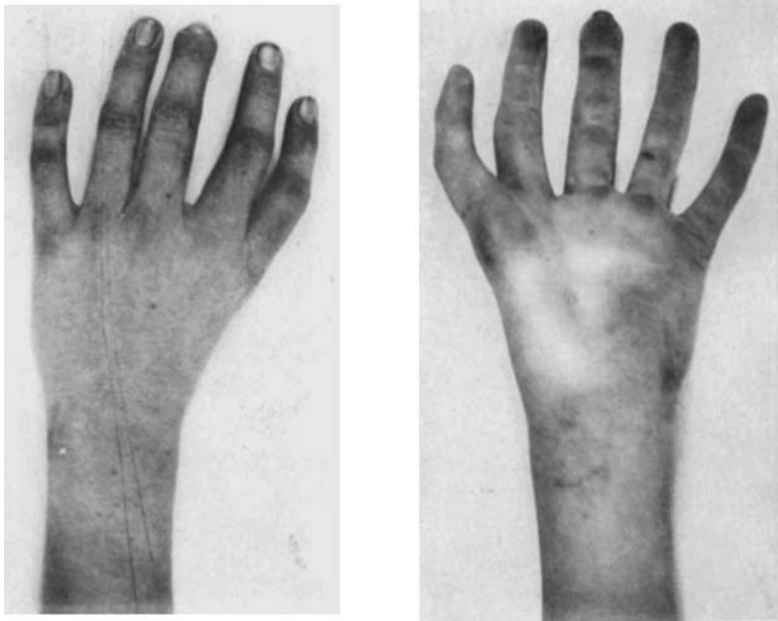


Fig. 2 Right hand of father

(fig. 1) hand, but this may be due to the fact that in figure 2 the hand was not held as flatly on the plate as in figure 1.

The son, the subject of this report is 13 years of age, 5 feet,  $8\frac{1}{2}$  inches in height and weighs 130 pounds. The photographs (figs. 3 and 4) show the marked deformity in the general appearance of the left hand and (figs. 5 and 6) the lesser deformity in the right. The II-V fingers are developmentally quite

normal for the middle finger of the left hand was injured in a recent accident. There is no webbing of the fingers and with the exception of the second and fourth fingers the sulci are normal. On the palmar surface of the fourth finger there are two sulci about 1 cm. apart opposite the first interphalangeal joint. The index finger has a similar peculiarity. The thumb has the appearance of the little finger of either hand. It would indeed be im-

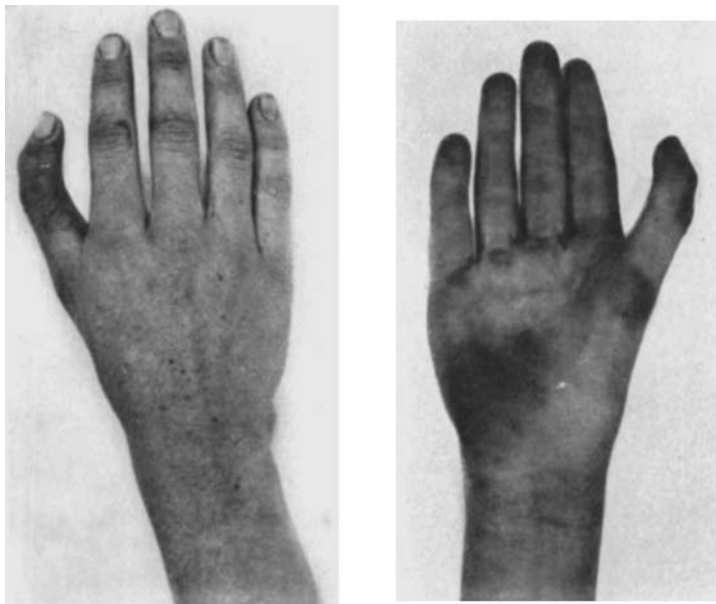


Figs. 3 and 4 Left hand of son

possible to identify it by means of a photograph of these digits alone, in the case of the left hand. The terminal phalanx and nail of the thumb are shaped exactly like the terminal phalanx and the nail of the little finger.

On the right hand the nail of the thumb, although smaller, is shaped more nearly like the normal thumb nail, and the terminal phalanx is also shorter and heavier than the terminal phalanx of the little finger. In each instance the thumb is 0.7

cm. longer than the little finger and markedly concave medially. The space between the index and middle fingers of the left hand is greater than normal and the index finger is concave medially. This is probably due mostly to the fact that the boy usually grasps objects between the index and middle fingers, rather than between the thumb and index fingers, because of the abnormal development and relation of the thumb. The thumb



Figs. 5 and 6 Right hand of son

of the right hand is also concave medially and small objects, such as a coin, when grasped by the right hand are placed between the dorsal surface of the terminal phalanx of the thumb and the tips of the fingers.

The thenar eminences are absent with apparently an absence of all the thenar muscles (figs. 4 and 6). Even the adductors appear to be absent and these facts no doubt account for the peculiarity in mobility. There is a lack of mobility of the metacarpals of the thumbs which is very noticeable because it limits

the motion of the thumbs to the movements possible in the other fingers.

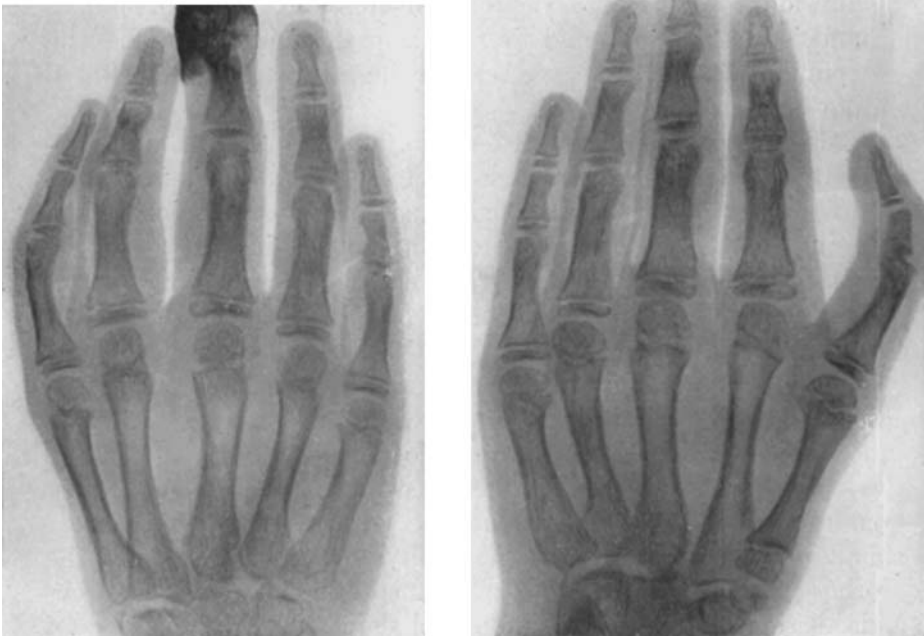
The position of the thumb in relation to the other fingers gives the forearm the appearance of being much longer than normal even though measurements show it not to be exceptionally long for a boy of his height. The hand is markedly widened at the metacarpophalangeal articulations. The left hand is 9 cm. wide. The width of the four fingers being 8 cm. The right hand is 9.3 cm. wide and the width of the four fingers 8.4 cm. A similar difference was present in the father's hands as stated above.

The photograph in figure 4 shows the smooth, glistening appearance of the skin in the palm of the hand. This is characteristic of both hands although a slight movement during the taking of the picture prevented its showing in figure 6. The skin seems to be drawn tightly so that very few sulci appear.

In the x-ray photographs (figs. 7 and 10) it is shown that the thumb of the left hand has three, well-formed, phalanges which in large part accounts for its length. In this case the age is such that the epiphyses show exceptionally well. There is nothing to indicate that the middle phalanx of the left thumb is rudimentary and will later fuse with the terminal phalanx. Indeed the middle is longer than the terminal phalanx and has a well-formed, normally-located epiphysis (fig. 7). Each of the other phalanges has also a well-formed epiphysis and seems entirely separate from the middle phalanx. The metacarpal of the thumb on this, the left hand, also is of interest for its form and length seem more nearly like a true metacarpal than the corresponding bone in a normal hand. Besides, the epiphysis is at the distal instead of the proximal end of the bone, the same as the epiphyses on the metacarpals of the other fingers (figs. 7 and 10). The metacarpal of the thumb is relatively long. The x-ray photographs were taken with the hands lying directly on the plate and with the machine over the same part of each hand. Measuring the negative we find that the metacarpal of the thumb is 6.7 cm. long while the metacarpal of the little finger is only 6 cm. long. The circumference of the first meta-

carpal also seems less than the circumference of the corresponding bone of the little finger. Normally the metacarpal of the thumb is shorter and thicker than any of the other metacarpals.

The x-ray photograph of the right hand is in some respects more interesting than that of the left. The extra phalanx of this thumb is not so well formed as that on the other thumb or



Figs. 7 and 8 Left and right hands of son

those of the other corresponding phalanges but nevertheless as shown in figure 9 it is a separate bone with a distinct diaphysis and epiphysis, articular surfaces and base and head. Besides showing the same peculiarities in regard to the extra phalanx, the metacarpal of this thumb shows a double epiphysis (figs. 8 and 11).

In all of the cases of three-jointed thumbs reported, heretofore, the middle phalanx is considered the extra phalanx.

Pfitzner, '90 who has done more work than anyone else on the development of the extremities, has reported several cases of three-jointed thumbs, but in all of these cases the three joints were only temporary for the extra, middle phalanx later fused with the end phalanx to form one bone. In no case did Pfitzner find an extra phalanx that had a distinct epiphysis, articular surface, head, and which in general had the shape of a normally developed phalanx as in the present case.



Fig. 9 Thumb of right hand taken laterally

Rieder '00, reported a family, most of whom had three-jointed thumbs. The father and four of the children had an extra phalanx in the thumb. Rieder claims that his cases differ from those reported by Pfitzner in that each end phalanx did have a distinct epiphysis entirely separate from the middle phalanx.

Dwight '07 in his report on the variations of the bones of the hand and foot considers the presence of an extra phalanx in the thumb. He says that in no case reported did the extra phalanx have an epiphysis in those of an age at which it could be expected to be present. He supports Pfitzner's theory that the extra phalanx is later fused with the terminal phalanx.

Salzar '98, also reported a case of a three-jointed thumb on each hand. In each instance the second phalanx was short and apparently rudimentary. The so-called pseudo-epiphysis shown in figures 8 and 11 is distinct and has the characteristics of a normal epiphysis. It would be hard to consider one a secondary epiphysis as has been done in most of the cases heretofore reported and in this case it seems quite evident that the first metacarpal had two true epiphyses. The presence of an extra epiphysis, however, has not caused any increased length as might be supposed, for although this metacarpal is longer than the metacarpal of the fifth finger of the same hand, it is not as long as the metacarpal of the thumb on the left hand, which has only one epiphysis. I am lead to believe that both epiphysis are true epiphyses, by comparing the x-ray photographs taken nine months apart. In figure 11, taken in November of 1916, both epiphyses have the appearance of true epiphyses as much as they did in the photograph taken in March of 1916 (fig. 8) or almost a year before. Hence although the supernumerary or cephalic epiphysis may fuse earlier than the basal or normal one it is clear that the former does not fall under what Thomson '06 speaks of as "scale-like epiphyses on the head of the first metacarpal which makes its appearance about eight or ten, and rapidly unites with the head." Since the first metacarpal in the left hand of this case, has a distally located epiphysis an interpretation of the latter in Thomson's sense would leave this metacarpal without an epiphysis.

Freund '05 also reported a case in which the middle phalanx had an epiphysis at the distal as well as at the proximal end. He compares his case to those reported by Pfitzner '00, and arrived at the same conclusion regarding the spurious character of these epiphyses. Pfitzner bases his conclusion upon the variation in ossification found in many cases which came under his observation. He thinks that in these cases of apparent supernumerary epiphyses the oestoclasts, instead of breaking down the epiphyseal cartilage in a straight-line front, extend into it in the form of processes or projections and thus produce an appearance which roughly simulates an epiphyseal line.

But as the photographs (figs. 8 and 11) show the breaking down of the cartilage in this case is being carried on in a straight line front. The portion of cartilage between the diaphysis and the extra epiphysis is as even and regular as in any normally developing bone. Hence Pfitzner's explanation does not apply here and this epiphysis seems to be a true and not a pseudo-epiphysis.



Figs. 10 and 11 Same hands about one year later

The mortising of the bases of some of the metacarpals is quite pronounced. On the medial side of the basal end of the third left metacarpal, for example, there is a deep notch for the articulation of the fourth metacarpal (fig. 10). There doesn't seem to be any explanation for the presence of this unusually deep notch.

Scrutiny of figures 10 and 11 will show that the carpal bones in the right and left hand vary both as to shape and relative position. In (fig. 10) of the left hand the greater multangular is considerably larger and completely overshadows the lesser multangular bone. In the right hand (fig. 11) the lesser multangular is larger than the greater multangular but the greater multangular is more laterally placed. Figure 10 also shows the triquetral bone to have a rectangular shape. Indeed, it looks not unlike a short phalanx. The triquetral bone shown in figure 11 is triangular and considerably larger than that in figure 10.

The thumbs in this case especially the left seem to be like a little finger in most respects except their position on the hand. Their movements are limited to the normal movements of a finger; they have the same number of phalanges as the other fingers: the nails resemble the nails of other fingers rather than thumb nails and their metacarpals have the characteristics of the metacarpals of fingers rather than those of normal thumbs. I do not think that the extra phalanx in either case can be considered as rudimentary because of the perfect development of each individual phalanx, the presence of a distinct normally-located epiphysis and the absence of any indication of fusion.

Professor Meyer to whom I am indebted for suggestions and assistance has offered a possible explanation for the occurrence of such a finger or thumb. If I understood him correctly he thinks that the apparent absence of the thenar muscles may indicate that the normal impulse to development of the thumb was absent and that hence the phalanges and also the metacarpals of the thumbs, assumed the form and relations characteristic of the fingers rather than of the thumb thus accounting for the extra phalanges in the thumb, the epiphyses in the supernumerary phalanges and also for the supernumerary epiphyses



Fig. 12 Normal left hand of a boy of approximately the same age, height and weight for comparison

in the metacarpal of the right thumb as well as the distally-placed, single epiphysis in the metacarpal of the left thumb.

Although Professor Meyer's suggestions seem to account for the results found in this case, he feels that we can not be certain that the thenar muscles are entirely absent. However, as far as can be determined from external appearances and from the limited mobility of the thumb they must be exceedingly rudimentary indeed if present.

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