FROM THE ORTHOPEDIC CLINIC OF THE »KAROLINSKA INSTITUTET .
(SENIOR SURGEON: PROFESSOR PATRIK HAGLUND).

ON CONGENITAL ULNAR DEVIATION OF THE FINGERS OF FAMILIAL OCCURRENCE
(„déviation des doigts en coup de vent")

BY

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(Translated from swedish manuscript).

In the following I shall describe a couple of cases of congenital ulnar deviation of the fingers. This deformity has previously only been described twice in the literature, viz. by *Boix* (1897) and by *I. Boerema* (1931).

Boix reports a case of congenital ulnar deviation of the fingers in a man, aged 50, who besides this deformity also displayed flexion contracture in the metacarpophalangeal and interphalangeal joints of the fingers. The patient's son, who died at the age of 2 years, was said to have had fingers resembling those of his father. At his birth his hands were so firmly clenched that they could only be straightened out with difficulty.

Boerema reports two cases. The first is that of a boy, aged 12, in whom a faulty position of the four ulnar fingers with simultaneous moderate flexion position of the metacarpophalangeal and interphalangeal joints was discovered when he was 2 years old. The second case was that of a boy, aged 13, in whom a deviation of the fingers completely corresponding to that described by *Boix* had been observed since his birth. The latter patient was treated with correcting hand splints, but skiagrams three years afterwards show that the deformity nevertheless has continued to develope.

I shall now proceed to describe cases of finger deviations in a woman, aged 48 (Mrs. H.) and her son, aged 12, (Åke H.) which correspond completely to the cases referred to above.

Mrs. H., case record No. 23495, has had an ulnar deviation of the fingers of both hands since birth (Fig. 1). She has never sought medical advice for the deformity. Otherwise she is in completely good health.

Present condition:— The hands, which are symmetrical, display a flexed position in the metacarpophalangeal joints of the index finger and the ring finger of about 25° in the case of the

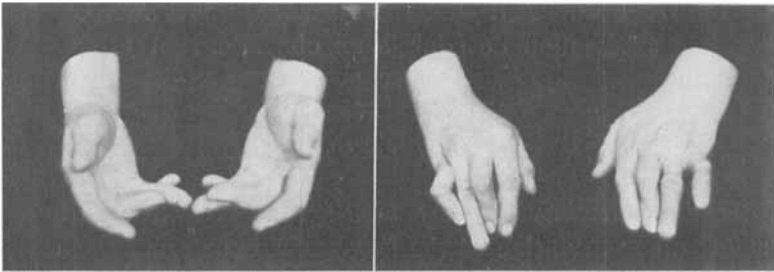

Fig. 1.

former and 45° in the case of the latter. The corresponding joint of the middle finger displays a normal position, but that of the little finger displays an overextension of 40°. The thumb is in a position of adduction. The interphalangeal joint of the index finger is in a flexed position of 10° and that of the middle finger is in a flexed position of 40°. The corresponding joint of the ring finger is somewhat overextended and that of the little finger is in a flexed position of 90°. The terminal phalanx of the index finger is in a normal position but that of the middle finger and that of the little finger are in a somewhat overextended position. The terminal phalanx of the ring finger is slightly flexed.

In the metacarpophalangeal joints the four ulnar fingers are in ulnar deviation, which in the joint of the index finger amounts to about 30°, in the joint of the middle finger to 35°, in the joint of the ring finger to 25°, and in the joint of the little finger to 45°.

The extensor tendon of the little finger is seen as a cord under the skin.

On the vola manus, which is shaped like a scoop, the skin is thin, tight and smooth. The mobility of the fingers is good. The gross muscular power is normal.

X-ray: (Fig. 2). The metacarpal bones occupy the normal position and their axis displays no ulnar deviation. But their

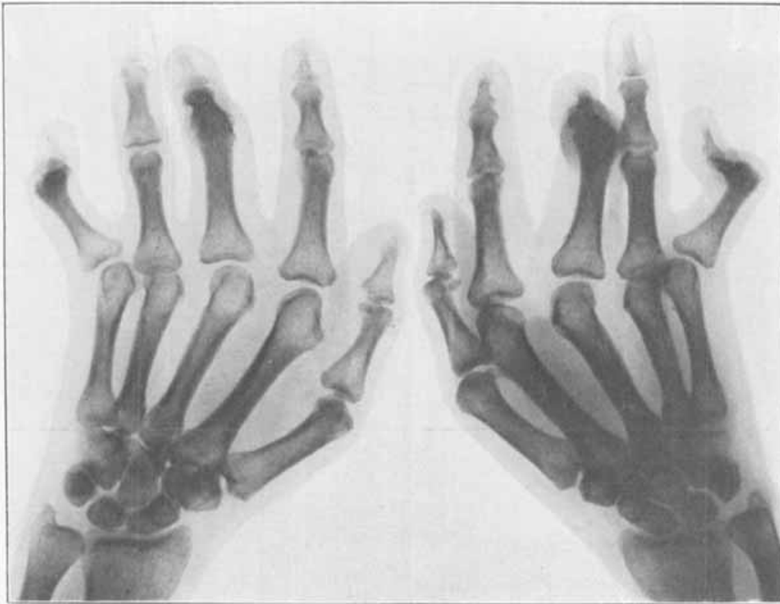

Fig. 2.

distal epiphyses deviate to the ulnar side, and the articular cavities between them and the basal phalanges of the fingers run obliquely from the ulnar side proximally to the radial side distally. The basal phalanges of the little fingers are in a position of marked subluxation.

Åke H., case record No. 19632. Fig. 3. According to his mother's statement the boy was born with firmly clenched hands. The thumbs were adducted and pressed so hard against the index and middle fingers that the impression of the thumbs could be

seen for a long period. The delivery was a forceps delivery after breech presentation. There were no complications.

When the boy was three years old he was treated by a doctor for pedes calcaneo-valgi congeniti. At the same time his father made splints for his hands according to the doctor's instructions.

Present condition:— The hands are symmetrical. The fingers are flexed in the metacarpophalangeal joints. The index finger is thus in a flexed position of about 45°, this flexion increasing

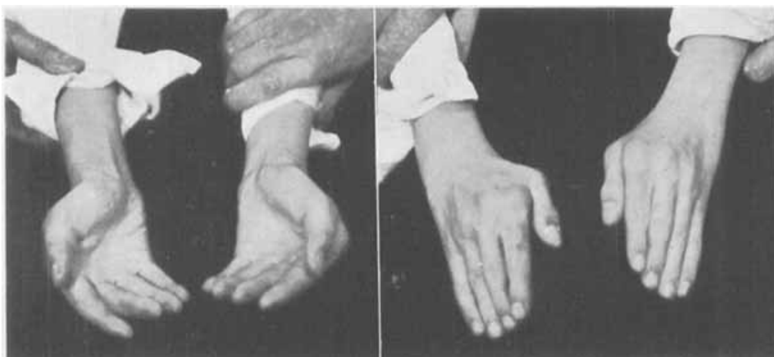

Fig. 3.

as far as the other fingers are concerned and being most marked in the case of the little finger (60°). The interphalangeal joints of the two ulnar fingers are in a position of extension, but the corresponding joints of the IInd and IIIrd fingers are somewhat flexed. The terminal phalanges are somewhat flexed with the exception of that of the ringfinger. The boy's hands, like his mother's, are bowl-shaped and look as if they had been formed round a small ball which had been lying in the vola manus. The skin of the vola manus is thin, tight, and smooth. The fingers can be corrected. The gross muscular power is good. The patient draws and writes well. The treatment consists of correcting hand splints (Fig. 4).

X-ray (Fig. 5):— The metacarpal bones do not deviate to the ulnar side. In the distal ends of the diaphyses of the IInd,

IIIrd, and IVth metacarpal bones faint indications of ulnar deviation are seen. Otherwise the bones of the hand are normal.

Mrs. H. and her son thus had the same congenital deformity of the hands. Even Mrs. H.'s mother is said to have had a faulty position of the fingers of one hand, but it was considered as engendered by an injury during threshing. Mrs. H.'s elder brother has the same deformity of the fingers as she herselt and besides very marked deformities of the feet, and the younger

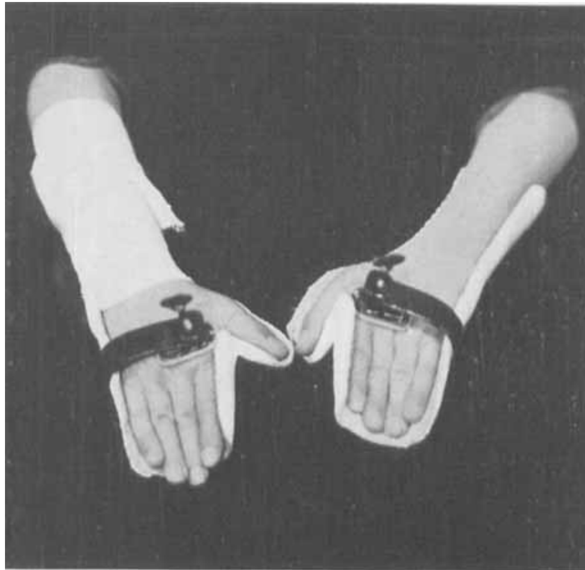

Fig. 4.

brother is said to have the same deformity of the hands. It is, however, impossible to obtain further information about them.

It is not known what the cause of this faulty position of the fingers, termed »déviation des doigts en coup de vent« by *Brissaud* may be.

Similar but acquired deformities have previously been described. The most well-known cause of the occurrence of such a faulty position of the fingers is doubtless chronic polyarthritis (*Hoffmann, Charcot, Hartman, Kahlmeter*).

Similar deformed hands are also seen among certain workers,

such as gardeners, blacksmiths, and milkmaids. *Fick* calls them workers' hands.

Billich states that in 40 per cent of the boys who after termination of their school years took up joinering, the fingers gradually deviated more and more to the ulnar side in the course of 10 years. In 50 per cent. of these cases the deviation exceeded 25°. Grasping the handle of the spade and the maul

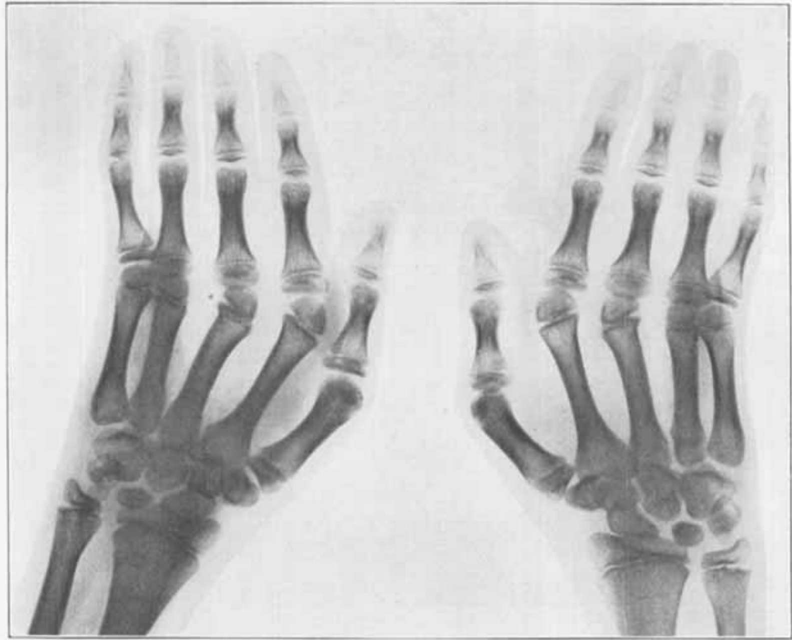

Fig. 5.

and the joiner's plane is said gradually to force the fingers into the faulty position.

Dittrich has observed a similar ulnar deviation of the fingers even in the small children and explains the occurrence through the continued grasping movements.

Feindel has described a case of »déviation des doigts en coup de vent« that occurred after injury to the ulnar nerve.

Boix supposes that the congenital faulty position of the fingers is due to the fact that the flexors during fœtal life have

commenced to exercise their effect before the extensors and that the former muscles owing to the position of their tendons have a tendency to draw the fingers towards the ulnar side.

Boerema supposes that the cause should be sought in an alteration during ossification in the distal metacarpal epiphysis of primary occurrence, the cause thus lying in the fœtal cartilage itself.

Jorge describes a congenital flexion contracture of the fingers in a mother and her three years old girl. He supposes, however, that the cause is a »rétraction palmaire congénitale«. It

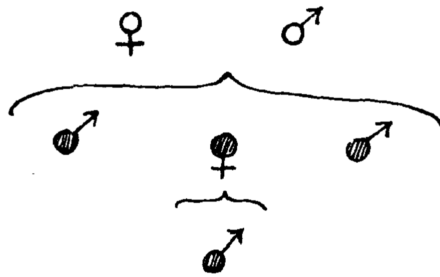

Fig. 6.

is thus only a question of a flexion contracture and not of ulnar deviation.

As to the differential diagnosis concerning the cases reported here it will mainly be the following groups of disorders that have to be considered:—

1. Rheumatoid polyarthritis.
2. Injury to nerves.
3. Dupuytren's contracture.

1. The following facts speak in favour of precluding rheumatoid arthritis.

a. Absence of contracture of the flexor tendons.
b. Typical skiagram, showing that there are no articular changes.
c. Familial occurrence.
d. Absence of any known rheumatoid infection.

2. Functional testing of the nerves has shown that injury to the nerves may be precluded.

3. There is certain resemblance to Dupuytren's contracture in the case of Mrs. H.'s hands. That Dupuytren's contracture nevertheless is not present in either of the cases reported here will be seen if we compare them with the condition in the said disease. The flexion which in Dupuytren's contracture ordinarily is seen in the first interphalangeal joint is completely absent here. In the mother there is moreover an overextension in the metacarpophalangeal joint of the little finger in contradistinction to the flexion which in Dupuytren's contracture otherwise is most marked here. Moreover the disorder is congenital in this case and is characterized by the marked, ulnar deviation of the fingers and the thin atrophic skin of the vola manus, no chords being palpable there.

As mentioned above, the cause of this congenital deviation of the fingers of familial occurrence is not known.

Billich has demonstrated that even in the normal individual there is an ulnar deviation of the index finger, when seen in relation to the longitudinal axis of the metacarpal bone. The middle finger occupies almost exactly the same axis as the third metacarpal bone. The two ulnar fingers show a slight deviation towards the radial side, whilst the index finger deviates 20° to the ulnar side. In this manner it is explained that it is just the index finger that deviates most in ulnar deviation of all the fingers.

It appears from the condition of our cases that the palmar aponeurosis is atrophic and shrivelled. This condition can of course engender a flexion contracture but not an ulnar deviation, as the palmar aponeurosis gives off four narrow projections which end the fibrous tendon sheaths of the long flexors of the fingers. These projections have about the same direction as the metacarpal bones.

The index finger is normally drawn towards the ulnar side by one of the m. interossei interni, the IIIrd and the IVth fingers each by its own m. interosseus internus. The Vth finger has its own abductor. All these muscles and their antagonists are innervated by the ulnar nerve and consequently it is not

probable that a preponderance of these small muscles should be able to engender an ulnar deviation.

The state of the long muscles of the fingers cannot explain the occurrence of the ulnar deviation either. *Billich* writes:

> »Eine Erklärung des ulnarwärts gerichteten Zuges durch die am Unterarm fast ganz von der Ellenseite entspringenden Flexoren — wie sie W. Braune herangezogen hat — kann auch kaum ausreichend sein, zumal diese einerseits durch ihre Einlagerung in den osteofibrösen Kanal des Ligamentum carpi transversum in ihrer ganzen Angriffsrichtung der Längsachse des Unterarmes sehr weit genähert werden, anderseits der Antagonismus der radialwärts ziehenden Extensoren einen Ausgleich bringen müsste«.

During intrauterine life the hand is nearly always clenched. This can explain a flexion contracture (*Derscheid-Delcourt*) but not the ulnar deviation. It is possible that the great pressure of the thumb on the radial side of the index finger can act in that direction. Åke H.'s mother in fact states that it was observed how hard the thumb was pressed against the index finger when the boy was born.

Just as the congenital flexion contracture according to *Dubreuil-Chambardel* is aggravated until complete ossification of the epiphyses, one of the cases described by *Boerema* was aggravated according to his statement. It is possible that high degrees of deviation can be avoided by letting the patients have correcting splints, to be worn at fixed times during day and night until the ossification is complete.

SUMMARY

In this paper an account is given of some cases of congenital ulnar deviation of the fingers of familial occurrence, viz. in a woman and her son, aged 12. Three similar cases have previously been described by *Boix* in 1897 and *Boerema* in 1931.

The deformity consists partly of a flexion contracture of all the fingers increasing towards the little finger, partly in an ulnar deviation of all the fingers. Moreover the vola manus is bowl-shaped.

From a differential diagnostic viewpoint the cases have been compared with Dupuytren's contracture, rheumatoid polyarthritis, and injury to nerves, but it has appeared that it is not any of these disorders.

The etiology has been discussed and several theories have been set forth by *Boix* and *Boerema,* e. g. a preponderance of the flexors of the fingers existing during fœtal life, or an change in the ossification of the distal metacarpal epiphysis of primary occurrence. It is emphasized that the preponderance of the flexors cannot possibly engender an ulnar deviation, but only a mere flexion position of the fingers. The writer mentions the possibility that the thumb through flexion and adduction when the hand during fœtal life is clenched may be able to influence the deviation in the ulnar direction. With regard to therapy, *Boerema* states that despite treatment with splints one of his cases was aggravated. The writer believes that continual use of splints until definitive ossification has occurred is capable of preventing too marked deformity. The splints made for the boy referred to are reproduced.

The paper is illustrated with photos and skiagrams.

In the bibliography works by *Billich, Boerema, Boix, Dittrich, Dubreuil-Chambardel, Feindel, Fick, Jorge,* and *Kahlmeter* are included.

RÉSUMÉ

L'article ci-dessus rapporte quelques cas de déviation ulnaire congénitale des doigts. Ces cas ont le caractère d'une anomalie familiale, se présentant chez une femme et chez son fils âgé de 12 ans. Trois cas analogues figurent déjà dans la littérature, rapportés par M. *Boix* en 1897 et par M. *Boerema* en 1931.

La déformité consiste d'une part en une contracture flexionnelle de tous les doigts, augmentant du côté du petit doigt, d'au-

tre part en une déviation ulnaire de tous les doigts. En outre la vola manus est de forme concave.

Au point de vue du diagnostic différentiel on a groupé ces cas avec la contracture de Dupuytren, avec la polyarthrite rhumatique et avec certaines affections nerveuses; mais l'auteur a pu constater que la déformité étudiée n'a aucun rapport avec ces maladies.

L'étiologie a été discutée par MM. *Boix* et *Boerema* qui ont avancé plusieurs théories; ainsi la naissance de l'affection a été attribuée à une prépondérance, dans la vie fœtale, de la traction des muscles fléchisseurs, ou bien à un trouble primaire de l'ossification dans l'épiphyse distale des os métacarpiens. Par contre, on a formulé l'objection que voici: la prépondérance des fléchisseurs n'aurait pas pu déterminer une déviation ulnaire, mais bien une position fléchie des doigts.

L'auteur indique une autre possibilité: le poing du fœtus étant tenu fermé, la pression du pouce aurait pu amener une déviation ulnaire des autres doigts.

ZUSAMMENFASSUNG

In diesem Artikel werden einige Fälle von angeborener Ulnardeviation der Finger mit familiärem Auftreten nämlich bei einer Frau und ihrem 12jährg. Sohn mitgeteilt. Drei entsprechende Fälle sind vorher — von *Boix* 1877 und *Boerema* 1931 — beschrieben.

Die Deformität besteht teils in einer Flexionskontraktur sämtlicher Finger gegen den kleinen Finger zunehmend, teils in einer Ulnardeviation aller Finger. Vola manus ist ausserdem schüsselförmig.

Differentialdiagnostisch sind die Fälle mit Dupuytrens Kontraktur, Polyarthritis rheumatica und Nervenaffektion verglichen worden, aber es hat sich gezeigt, dass das Leiden mit keiner dieser Krankheiten in Verbindung steht.

Die Aetiologie ist besprochen worden, und viele Theorien wurden von *Boerema* aufgestellt, z. B. wurde das Entstehen des

Leidens einem vorhandenen Uebergewicht des Zuges der Flexoren im Foetalleben oder einer primär auftretenden Ossifikationsstörung in der distalen Epifyse der Metatarsalknochen zugeschrieben.

Dagegen ist hervorgehoben worden, dass ein Flexionsübergewicht keine Ulnardeviation, aber sehr wohl eine Flexionsstellung der Finger hervorrufen können sollte. Der Verfasser betont die Möglichkeit, dass der Daumen, wenn die Hand während des Foetallebens als Faust gehalten wird, eine ulnare Abbeugung der übrigen Finger mitsichführen können sollte.

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