INCIDENCE OF NEONATAL HIP INSTABILITY : ARE THERE SEASONAL VARIATIONS ?

J. K. ANAND[1], I. MODEN[2], J. W. MYLES[3]

We wanted to establish whether the season of birth affected the incidence of neonatally diagnosable hip instability in Eastern England. Data relating the numbers of maternities to the numbers of cases of congenital displacement of the hip diagnosed in neonates were analyzed month by month over a period of 8 years. The study covered 185,744 maternities and 154 cases of displaced hips during the period 1979-1986. While there were wide fluctuations in the monthly incidence, there was a striking excess of the malformation in late winter and early spring, i.e. in January, February, March and April, and a progressive decline in the succeeding months. The cause or causes of the seasonal variation are unknown.

Keywords : hip ; instability ; congenital ; seasonal ; incidence.
Mots-clés : hanche ; instabilité ; congénital ; saisonnier ; incidence.

INTRODUCTION

The question as to whether there is a significant seasonal variation in the incidence of congenital displacement of the hip has been reviewed by Tönnis (13) and addressed, among others, by Record and Edwards (11), Andrén and Palmén (1), Kupper *et al.* (8, 9), Weissman and Salama (14), Barlow (3), Woolf, Hoehn and Coleman (16), Wynne-Davies (17, 18), Mackenzie (10), and Artz *et al.* (2). These contributions offer conflicting results. Most of these studies include late-diagnosed congenital dislocation of the hip (CDH). We present data from the east of England over a period of 8 years during which a total of 185,744 women were delivered. Our data refer exclusively to hip instability diagnosed neonatally, and presumably resulting from factors operating in utero or as an expression of genetic load. A very definite seasonal incidence of congenital displacement of the hip is apparent.

MATERIALS AND METHODS

All live births, both hospital and domiciliary, in East Anglia are recorded, and all neonates are examined by medical staff. When the diagnosis of congenital displacement of the hip is made, this is recorded and coded (ICD code 754.3). The monthly data relating to 185,744 maternities and 154 cases of CDH diagnosed neonatally during the years 1979 to 1986 were studied. During this period, ultrasound had not been adopted for screening purposes. Only the Ortolani and Barlow techniques were in use.

RESULTS

No cases were recorded in the births occurring in September, November and December. January and February produced aggregates of 24 and 23 malformations respectively with the incidence rising to 34 in March and 31 in April. In May the incidence fell to 10 cases, and this level then

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persisted through the summer and then declined through the autumn to nil in November and December prior to a rise in January.

The monthly distribution of CDH and correlation with maternities is illustrated in the graph (fig. 1).

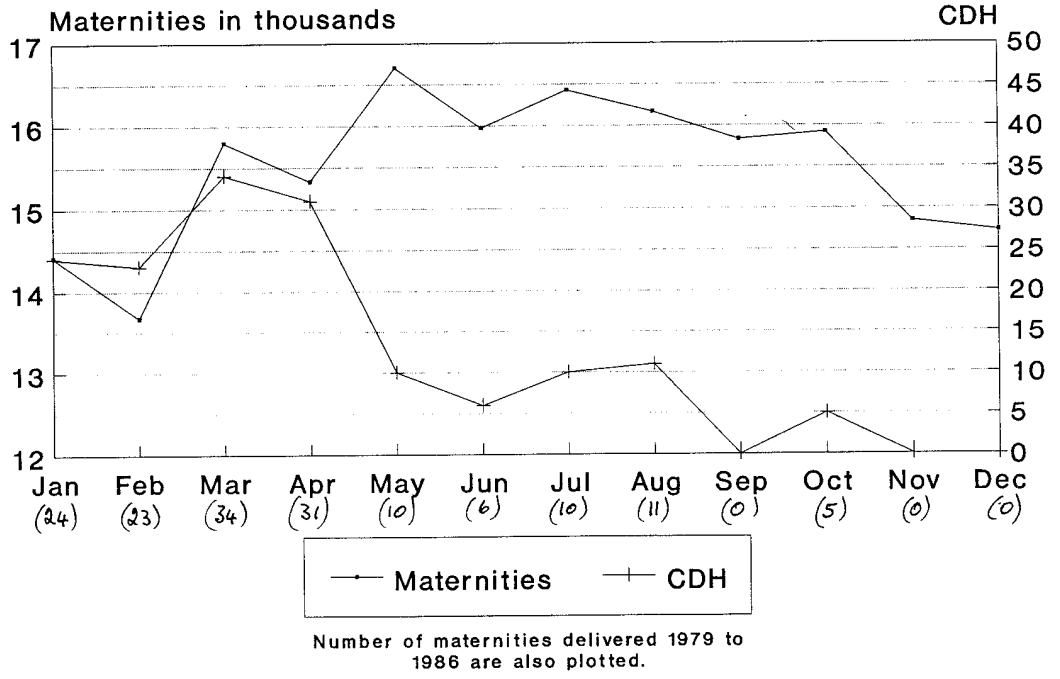

Fig. 1. — Graph showing the high incidence of early CDH in late winter and early spring months 1979 to 1986 inclusive.

DISCUSSION

Conditions which are coded under the name "congenital dislocation of the hip" may be true articular malformations, as indicated by a positive Ortolani or Barlow test, but some cases may be missed and false positives may be recorded. The aggregated data presented here should even out the vagaries of excessive enthusiasm by some examiners, lack of experience in others, genetic factors which may affect the incidence in some communities and also random variations. Late CDH is not included in this study.

Our records show a clear excess of neonatal CDH in the late winter and early spring maternities. This finding is compatible with that resulting from a large series from Birmingham (11). The Birmingham study however included late cases of congenital dislocation. Czeizel *et al.* (7) also found the maximum incidence in January. Their low was reached in May and June. The Swedish patients studied by Andrén and Palmén (1), 816 infants born in the years 1953 to 1960 and yielding a positive Roser-Ortolani test, and 1313 with CDH undetected at birth but mostly discovered after one year of age, showed a peak in September, October and November and a trough in March to May. From Oslo, Bjerkreim and Hagen (5) reported that late-diagnosed CDH reached the peak incidence in early October, while the month of April showed a low incidence. This Norwegian investigation found no obvious seasonal influence on the incidence of neonatal congenital dislocation. Our findings are quite the reverse. The Vikings of Scandinavia did colonize East Anglia but our population is very mixed — there were also the Saxons, the Belgae, the Celts and of course late arrivals from the rest of Europe and indeed the rest of the world. Kupper, Ackermann and Schmerling (8) from Halle-Wittenberg found, in 983

patients with congenital dislocation of the hip, a significant seasonal variation, with higher rates particularly in October and in November, but in a second sample of 333 neonates with a positive Ortolani sign, the authors concluded that "such a statement cannot be made". Barlow (3) working in northwest England found no seasonal variations. Wynne-Davies (17, 18) in Scotland found a winter/summer ratio of neonatal cases of CDH of 1/1.5. She found that in the children with late-diagnosed CDH the winter/summer ratio was 1/1.3 and suggests that in this latter group the genetic factor of acetabular dysplasia is of greater significance (17, 18). Mackenzie (10) analyzed the findings from a cohort of 76,675 neonates from the northeast of Scotland. He reported the highest incidence in the months of June, July and August, with the December, January and February quarter close behind. The spring months had the lowest incidence, while that in the autumn was similar to that in the summer quarter. Wilkinson (15) reported a smaller series but with a higher incidence of neonatal CDH than ours, namely 3.7 per 1000 live births compared with 0.83. His finding of a high incidence in February reflects an observation of a maximal incidence in March, and his second peak in September corresponds to our second peak in August. Weissman and Salama (14) in Israel found a preponderance of positive Ortolani tests on children born in winter. In New York, Artz *et al.* (2) conducted a large study which showed no seasonal trends. On the other hand Woolf and colleagues (16) in Utah discovered a preponderance of CDH in the autumn and early winter births.

Our data set probably represents a different genetic composite from most of those referred to above. The excess of late winter and early spring incidence is impressive and it is interesting that in the months of November and December the incidence was nil. It should be pointed out that seasonal temperatures, hours of sunshine and rainfall as well as other ecological and genetic influences vary between the countries and regions from which published data are available. Kupper and Ackermann (9) studied the monthly mean air temperature and the incidence of congenital dislocation of the hip for samples from Birmingham, Jerusalem, Budapest, Oslo and Halle. No clear relationship emerged. Andrén and Palmén (1) speculated that seasonal variations in levels of hormones such as estrogens and relaxin may influence the genesis of congenital dislocation of the hip. Certainly hormones do influence the laxity, in pregnancy, of ligamentous structures. Steinetz *et al.* (12) found that the ability of relaxin to relax and lengthen the interpubic ligament varied in different strains of mice according to the season. The relaxin reference standard lost 30-40% of its activity in the 3-month period of January to March. In harmony with the theory of hormonal influences on the capsular and ligamentous structures is also the finding by Berezin (4) of pelvic insufficiency varying in intensity according to the season.

Chen *et al.* (6) believe that the fourth, fifth and sixth months of intrauterine life are critical for the seasonal effects on the development of the hip joint, and that the freedom of movement of the limbs in this period must remain unimpeded if CDH is not to develop. Tönnis (13) has suggested that there may be seasonal variations in the production of amniotic fluid. He also suggests that seasonal influences may act through changes in the environment rather than through changes in the ambient temperature. It seems reasonable to postulate that the defect is multifactorial, the interplay of genetic factors and the intrauterine environment determining the condition at the time of birth. The seasonal changes during the pregnancy must in some way affect the intrauterine environment of the fetus.

CONCLUSION

In the East Anglian population studied in the period from 1979 to 1986, there was a clear seasonal influence on the incidence of neonatally diagnosable congenital hip instability. Late winter and early spring births showed a higher incidence of this malformation. How the changing seasons exert their influence remains unknown.

Temperature differences are unlikely to have a direct bearing on the genesis of this defect which is probably multifactorial.

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SAMENVATTING

J. K. ANAND, I. MODEN en J. W. MYLES. Frekwentie van de neonatale heupinstabiliteit ; zijn er jaargetijde gebonden variaties ?

De auteurs wilden vaststellen of het jaargetijde ten tijde van de geboorte van invloed was op de incidentie van, in de neonatale periode te diagnosticeren, heupinstabiliteit in Oost-Engeland.
Data, die het aantal bevallingen relateren aan het aantal gevallen van congenitale heupluxatie, gediagnosticeerd in neonaten, werden maandelijke geanalyseerd over een periode van 8 jaar.
Het onderzoek omvatte 185.744 bevallingen en 154 gevallen van congenitale heupluxatie gedurende de periode 1979-1986.
Terwijl er grote schommelingen waren in de maandelijkse incidentie, was er een opvallende toename in het aantal gevallen met deze malformatie tegen het einde van de winter en in de vroege lente, dat wil zeggen, in januari, februari, maart en april en een afname in de daarop volgende maanden.
De oorzaak of oorzaken van deze verschillen per jaargetijde zijn onbekend.

RÉSUMÉ

J. K. ANAND, I. MODEN et J. W. MYLES. Fréquence de l'instabilité de la hanche à la naissance. Existe-t-il des facteurs saisonniers ?

Les auteurs se sont demandé si l'époque de la naissance pourrait jouer un rôle dans la genèse d'une instabilité de hanche, dans l'est de l'Angleterre. Ils ont étudié des dossiers de luxation congénitale et d'instabilité de hanche, constatées à la naissance, mois par mois, pendant une période de 8 ans. Ils ont relevé 185.744 accouchements et 154 dysplasies de hanche entre 1979 et 1986. Ils relevèrent de larges variations dans la fréquence de ce diagnostic au cours des différents mois de l'année, mais il y avait une augmentation frappante de la malformation à la fin de l'hiver et au début du printemps, c'est-à-dire en janvier, février, mars et avril et une diminution progressive pendant les mois suivants. La où les causes de cette variation saisonnière restent inconnues.

TYPE II SYNDACTYLY OR SYNPOLYDACTYLY

L. DE SMET[1], G. FABRY[1]

A family with syndactyly type II or synpolydactyly is described. The autosomal dominant inheritance is confirmed by this pedigree. In combination of this anomaly a brachymesophalangia of the fifth finger was inherited by most family members. The duplicated phalanx was resected and the syndactyly separated in the proband with excellent functional and cosmetic results.

Keywords : synpolydactyly ; syndactyly type II.
Mots-clés : synpolydactylie ; syndactylie type II.

INTRODUCTION

Synpolydactyly is the second type of isolated syndactyly according to the classification of Tentamy and McKusick (9) (see table I). It must be distinguished from the polysyndactyly which is a preaxial polydactyly in combination with classical syndactyly of the third and fourth ray. The latter has to be considered as a polydactyly, since in pedigrees syndactyly only occurs in association with polydactyly. In synpolydactyly syndactyly can occur in the absence of polydactyly. Other authors have made a separate classification and called it central polydactyly. This is a pure morphological definition without any indication of genetics or teratology. The associated central polydactyly-bony syndactyly-typical cleft was demonstrated by the experimental and clinical work of Ogino (5).

Reported cases and/or pedigrees are sparse. In this paper we report a family with 3 affected members. In association with the synpolydactyly a brachymesophalangy of the fifth finger (type A_3 brachydactyly of Bell according to Tentamy and McKusick (9)) was transmitted to most members of this kindred.

Table I. — Classification of syndactyly
(adapted from Tentamy and McKusick (9))

Type A : isolated
zygodactyly (cutaneous long-ring)
synpolydactyly
ring-little finger
complete hand
including metacarpal synostosis
Type B : syndrome-related

CASE REPORT

The family pedigree is presented in fig. 1. The proband presented at the age of 3 years for treatment of a congenital syndactyly of the right hand.

The patient was a healthy first-born female child. The parents were unrelated ; the mother's age was 28 years, and pregnancy was uneventful. Psychomotor development was normal ; anthropomorphic data were between P50 and P90. A unilateral complete syndactyly between the third and fourth finger and a bilateral brachymesophalangy of the fifth finger were present (fig. 2). The same "crooked" little finger was present in the father's hands (fig. 3). Radiographs of both hands demonstrated an extra terminal phalanx. The middle phalanx of the fifth finger of both hands was small and deviated. The feet were clinically and radiologically normal. At operation the cu-

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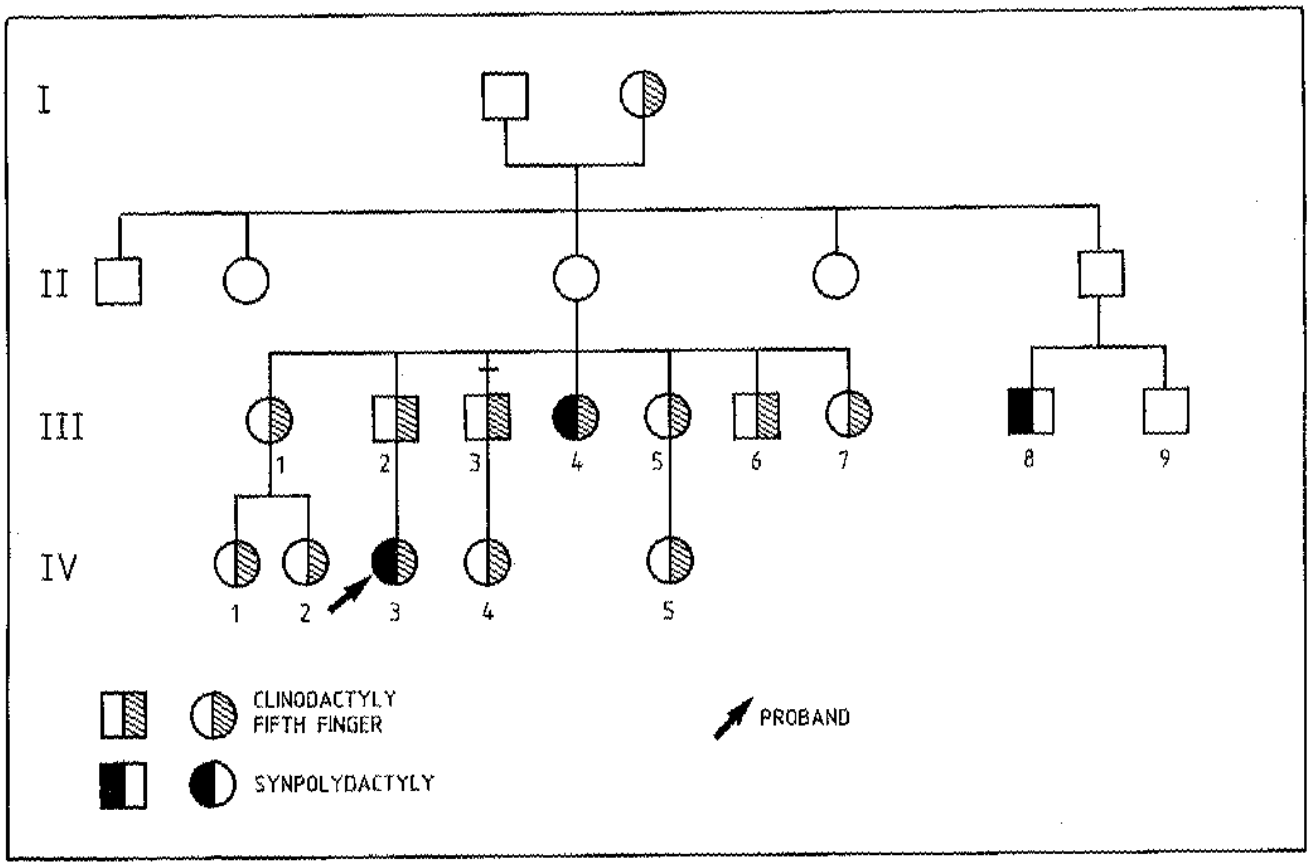

Fig. 1. — Pedigree of the kindred.

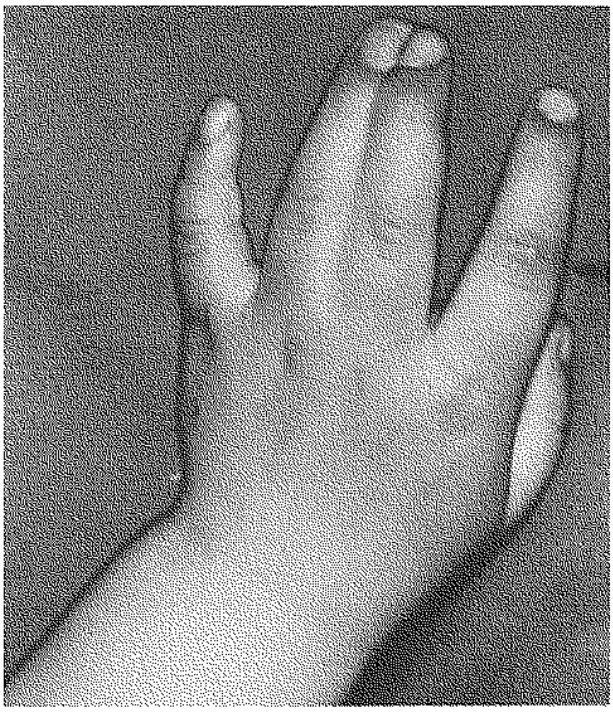

Fig. 2. — Proband's hand ; complete syndactyly of the third and fourth finger, synochia, clinodactyly of the fifth finger.

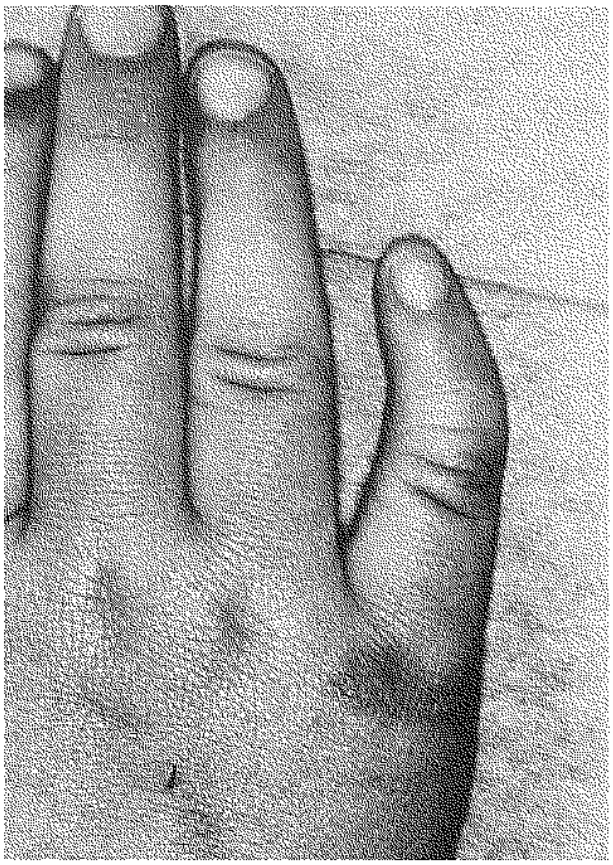

Fig. 3. — Father's hand with clinodactyly of the fifth finger.

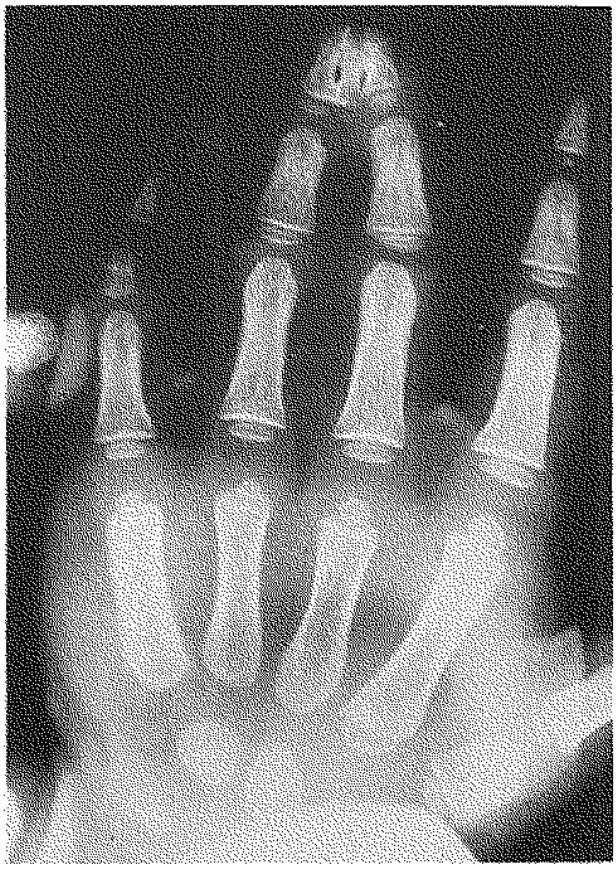

Fig. 4. — Radiograph of the proband's hand, demonstrating the extra terminal phalanx enclosed in the syndactyly.

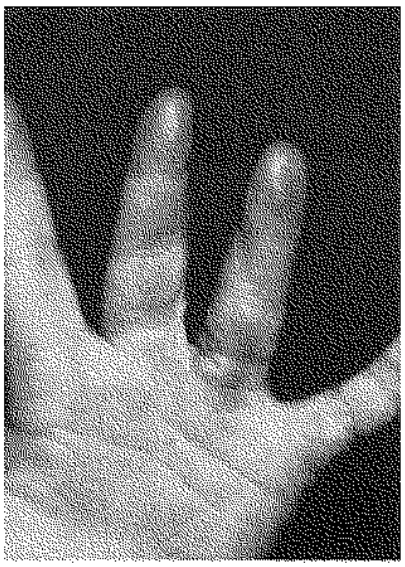

Fig. 5a

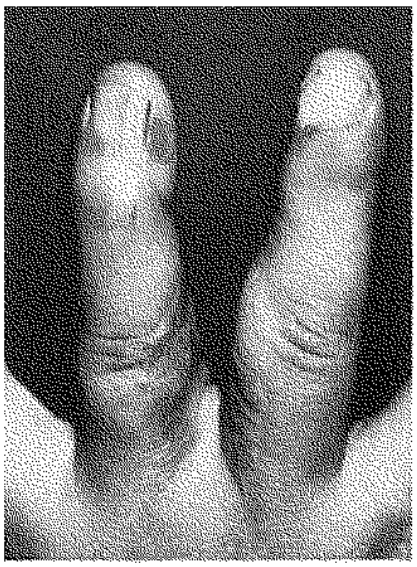

Fig. 5b

Fig. 5c

Fig. 5. — a, b, c : clinical result of the operative release operation.

taneous syndactyly was separated as well as the unique nail, according to Flatt's technique. The supernumerary phalanx was resected. A tight fibrous band was present between both fingers and had to be resected to permit full division and creation of the new web. Results after 1 year are very satisfying (fig. 5).

An aunt of the proband (III, 3) was examined for scar retraction of a previously operated syndactyly of the right hand. Preoperative radiographs revealed a similar terminal phalanx. The father's cousin (III, 8) has been treated elsewhere for a syndactyly ; no details were available. The crooked little finger was present in most family members, except for the proband's grandmother.

DISCUSSION

Syndactyly has been classified by several authors (7, 8, 9) as well as polydactyly. Synpolydactyly or the type II of Tentamy and McKusick (9) is one of the rare isolated, nonsyndromal syndactylies. Case and kindred reports are sparse (1, 3, 6, 8, 10). According to McKusick an autosomal dominant inheritance is the rule (2). In one of the larger series, published by Tada (8), no patient had a positive family history. It is hard to consider them all as new mutations, but since most patients were very young, we do not know if the trait was transmitted to other generations. As in all other cases the absence of major general anomalies is striking.

The association of the mesophalangia of the fifth finger can be considered a coincidence, since Miura (4) found a brachymesophalangia of the fifth finger in 56.7% of all middle-ring finger syndactylies. Since our case was particularly simple, treatment was not more complicated than a simple syndactyly release, and a good outcome was expected. It is clear that for such cases the pessimism of Wood cannot be shared (11, 12).

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SAMENVATTING

L. DE SMET, G. FABRY. Synpolydactylie of syndactylie type II.

Een familie met syndactylie type II of synpolydactylie wordt beschreven. De autosomaal dominante overerving wordt bevestigd. Tevens werd bij de meeste familieleden een brachymesophalangie van de vijfde vinger op dezelfde wijze overgeërfd. De spreiding van de syndactylie en de resectie van de ontdubbelde falanx hebben tot een uitstekend funktioneel en esthetisch resultaat geleid.

RÉSUMÉ

L. DE SMET, G. FABRY. Syndactylie type II ou synpolydactylie.

Les auteurs rapportent le cas d'une famille dont plusieurs membres présentent une syndactylie type II ou synpolydactylie. La transmission autosomale dominante est confirmée par cette généalogie. Une brachymesophalangie du cinquième doigt existait en outre chez plusieurs membres de cette famille. La séparation de la syndactylie et la résection de la phalange surnuméraire ont mené à un résultat fonctionnel et esthétique excellent.