Treatment of congenital syndactyly of the fingers

BACKGROUND Syndactyly or webbed fingers is one of the most common congenital malformations of the upper extremities, but it comprises few new cases annually. The purpose of treatment is to enhance hand function.

METHOD The article is based on current text books and literature searches in PubMed as well as the authors' clinical experience within this field.

RESULTS The purpose of surgical treatment is to separate the fingers and reconstruct a webspace. It is difficult to indicate exact treatment results because of large variations in the extent of the deformity. For syndactyly involving only soft tissue (simple syndactyly), a good functional result is achieved with a less than 10 % risk of complications. Syndactyly where also the bones have fused (complex syndactyly) or where there is additional bone formation between two digital rays (complicated syndactyly), gives a poorer functional outcome and a higher risk of complications. Gradual stretching of the tissue using a distraction device enables separation of fingers one was previously reluctant to separate.

INTERPRETATION It should be possible to expect safe separation with a good and independent function of the fingers with surgical treatment. The parents should be informed that the surgical treatment is a reconstructive procedure that may require secondary corrections.

Hebe Désirée Kvernmo
hebe.kvernmo@gmail.com
Section for Upper Extremity- and Microsurgery
Department of Orthopaedics
Oslo University Hospital

Jan-Ragnar Haugstvedt
Department of Orthopaedics
Østfold Hospital, Moss

MAIN POINTS

Syndactyly should be assessed during the examination of the newborn baby and noted on the birth report which is sent to the Medical Birth Registry

The child must be referred to a department with expertise in hand surgery as well as to the paediatric department and for genetic testing in case of associated conditions and anomalies

Proper attention paid to the parents and thorough information about the treatment are necessary to ensure a good result

Treatment of syndactyly is a reconstructive procedure, but safe separation should be expected with proper independent functioning of the fingers

The term *syndactyly* derives from the Greek *syn* (together), and *daktylos* (finger), and refers to the clinical appearance of fingers which have not undergone a normal process of separation in intrauterine life. There is a large degree of heterogeneity in the extent of the malformation, which may have functional, developmental and cosmetic implications (1, 2). In this article we wish to give an overview of the condition and how to deal with it.

Method

This article is based on current text books, participation at international conferences, the authors' clinical experience within this field as well as articles found through a selective literature search in PubMed using the MESH terms (syndactyly/surgery* OR syndactyly/methods*). The selection was restricted to long-term results of surgical treatment published in English-language journals in the period from 2000 until the search date 18.12. 2012. The articles dealing with results of surgery were all based on simple case series without randomisation or blinding.

Epidemiology

The Danish registry of congenital malformations showed an incidence of malformations of the upper extremities for the period 1984–93 of 14.6 per 10 000 live births (3). The incidence of syndactyly was 2.8 per 10 000. Data from England and Wales, by comparison, show that syndactyly occurs in one in every 2 400 live births (4). The condition represents approximately 20 % of all malformations of the hand (3, 5) and occurs twice as frequently in boys. 50 % of syndactylies are bilateral, and bilateral, simple syndactyly between the long finger and ring finger is most frequent. Involvement of the thumb and index finger, or the index finger and long finger is least frequent (6, 7), except when related to a syndrome (6). Figure 1 illustrates which webspace is involved (8). Complex syndactyly represents only 16.5 % of the cases (5).

Classification

The classification of syndactyly is shown in Figure 2. The term *simple* syndactyly is used if only the soft tissues are involved, and *complex* syndactyly if the bones of adjacent fingers are fused (synostosis) (1). The term *incomplete* syndactyly is used to describe a fusion that is proximal to the distal phalanx and *complete* if the syndactyly continues up to the distal phalanx. The term *complicated* syndactyly describes syndactyly with bone formation between two digital rays (hidden polydactyly), and *acrosyndactyly* describes syndactyly involving the distal phalanges, however with a proximal webspace.

Classification and aetiology

The development of the hand starts on day 27 of intrauterine life (9, 10). The fingers are normally separated between day 44–46. Most syndactylies appear in this period and can be attributed to errors in programmed cell death (apoptosis), which normally involutes the embryonic interdigital tissue and enables the formation of separate fingers (1, 10). This tissue normally involutes from distal to proximal and explains the phenomenon

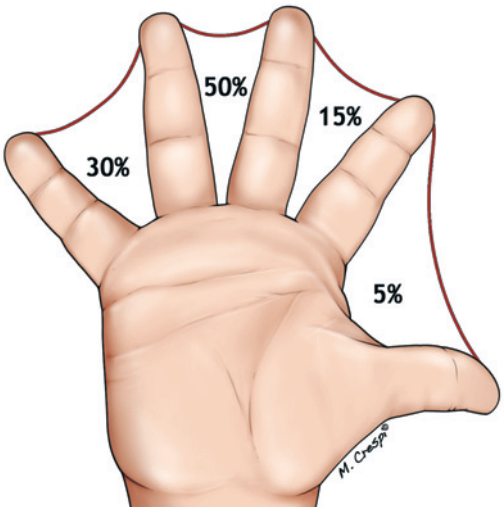

Figure 1 *Schematic representation of involved web spaces (8)*

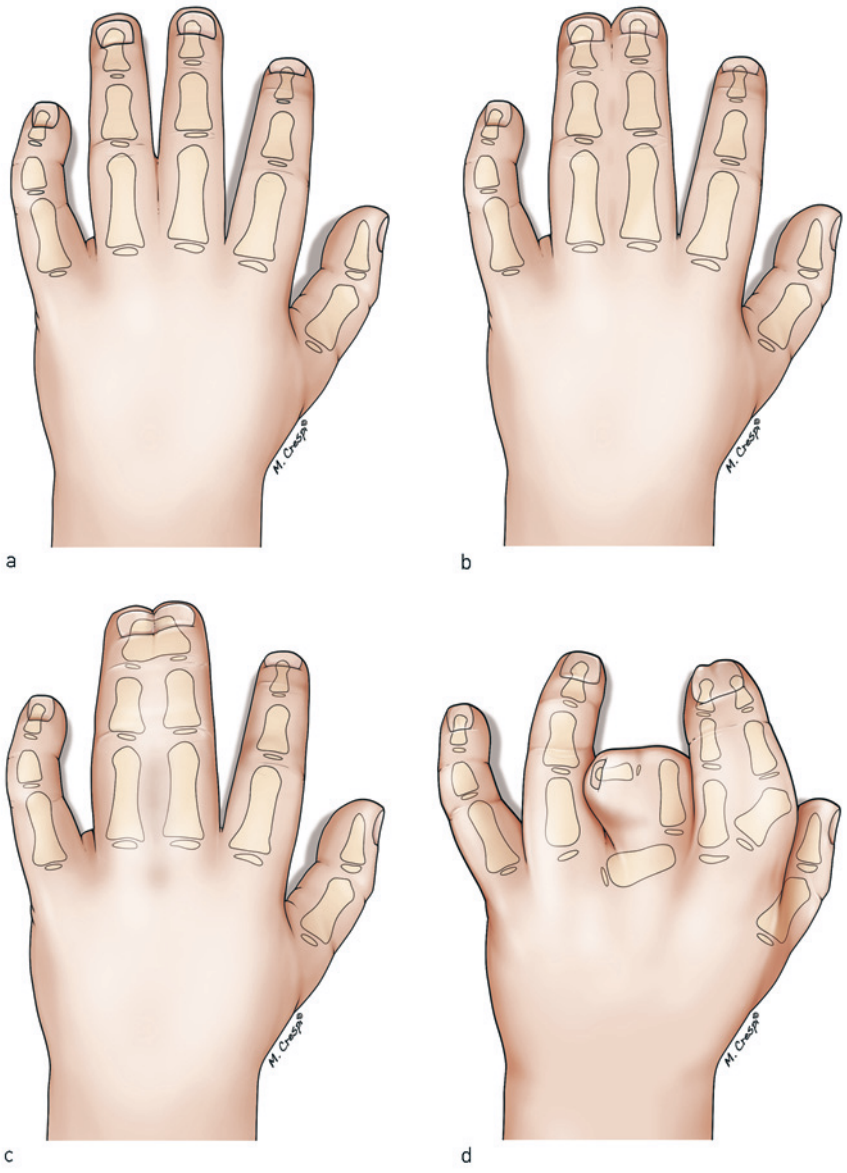

Figure 2 *Classification of syndactyly. Syndactyly between long and ring finger. a) Simple incomplete syndactyly b) Simple complete syndactyly c) Complex complete syndactyly d) Complicated syndactyly*

of simple incomplete syndactyly. The exception to the rule is acrosyndactyly, where the most common form is associated with amniotic band syndrome, which causes fingers to fuse after the separation process is completed (1). Differentiation of other organ systems proceeds in parallel with that of the hand, which explains associated anomalies in a number of syndactylies (2, 10). In 1976 the International Federation of Societies for Surgery of the Hand introduced a classification system according to which congenital malformations of the upper extremities are classified into seven categories (11). Syndactyly is placed in group II together with other failures in differentiation.

The majority of syndactylies occur isolated and with unknown cause. Factors that may have affected the intrauterine environment, such as exposure to teratogenic agents, virus infections or other diseases in early pregnancy, are postulated as possible causal hypotheses (2). 10–40 % of the cases have a positive family history (1, 2, 12). When syndactyly occurs isolated and there is a positive family history, the syndactyly has always an autosomal dominant inheritance (1). It is often both complex and complicated. However, the dominant gene shows reduced penetrance and variable expressivity, so that the condition varies greatly from one generation to the next (1). Syndactyly is moreover a feature of at least 28 syndromes. The most common are Apert syndrome, Poland syndrome, amniotic band syndrome and multiple craniofacial syndromes (1). Only 5 % of all malformations of the upper extremities are part of a defined syndrome (2).

General examination of newborns

When a child with syndactyly is seen, it must be remembered that other simultaneous malformations can occur due to chronological proximity to the intrauterine development of the hand (2, 10). The simple syndactylies are not, however, associated with other congenital malformations (2) and do not require further investigation than the normal assessment during the examination of the newborn baby. If it is the complex form and it is not inherited, discretion is exercised. In most cases fusion of the bones of two adjacent fingers will not entail further investigation apart from carrying out a clinical examination of the upper extremities, feet, head /face and thorax. If associated conditions are suspected, or the syndactyly is related to a syndrome, the child is referred to a paediatric department and to geneticists who assist in further investigation.

It is important to remember that all congenital malformations that are discovered at the maternity or neonatal unit are notifiable to the Medical Birth Registry. Mandatory

notification also applies to the less extensive conditions, such as simple incomplete syndactyly. When the diagnosis is unclear and the child is referred for further investigation, it is important to note this on the birth report. This enables the Medical Birth Registry to follow up on the diagnosis, resulting in as correct registry data as possible. Registry data based on an accurate medical history may also potentially help to reveal causal factors for a number of syndactylies.

The child should have an early referral to a department with expertise in hand surgery, out of regard for the parents' need for information, and planning and possible need for coordination of the further treatment.

The assessment for hand surgery

Clinical examination provides an overview of the extent of involved fingers. Closer clinical examination will make it possible to distinguish between simple, complex and complicated syndactyly. The presence of an active range of motion in the interphalangeal joints with well-defined flexion and extension creases indicates normal joint anatomy and simple syndactyly. If there are no flexion and extension creases and the joints are not mobile, complex or complicated syndactyly must be suspected. Lack of movement between the distal phalanges of two digital rays and fused nails indicate synostosis. In syndactyly associated with other clinical syndromes such as Poland syndrome, Apert syndrome or amniotic band syndrome, clinical examination of the upper extremities, feet, head/face and thorax is needed.

Plain X-ray examination of the affected hand should always be included in the primary assessment to reveal any synostoses, hidden polydactylies or other skeletal deformities, and is particularly important in complex syndactylies.

Assessment for surgical treatment

Surgery is recommended for most children (1, 8, 12–14). The purpose of treatment is to enhance the functional level. Emphasis is placed on the cosmetic aspect and the child's natural ability to adapt to the malformation. Separation of fingers is contraindicated if it can be seen that this could reduce existing functional level, for example if components of the fused fingers are missing, which makes it impossible to create independent, stable and mobile fingers (13). Medical conditions may likewise be a contraindication for surgery. Mild, incomplete syndactyly without functional limitations is only a relative indication for surgery.

Timing of surgery

While there are few controversies with regard to the *indication* for surgery, the *timing* of surgery is to some extent discretionary. The question to be asked is not how early the procedure can be carried out, but rather how long it can be postponed based on the functional needs of the hand. (1). The timing is assessed for individual cases and depends on which fingers are involved, as well as the extent and complexity. Likewise there may be other malformations which must be dealt with before the surgical treatment of the hand. When several procedures are required, these should be carried out before the child reaches school age.

Generally it can be said that simple syndactyly of the web between second and third fingers can safely be separated at the age of 1–2 years (1, 8, 12). Surgery on a larger hand reduces the risk of scar contracture and development of web creep, where the webbing creeps distally as the hand grows.

For syndactylies of the first and fourth interdigital webs and syndactylies with fusions of several fingers, surgery is recommended at the age of 4–9 months (1, 8, 12, 15). The ability to locate the hand in space and perform grip and pinch manoeuvres are cortical functions that develop before the age of one year. Coordination continues to develop until the age of three years (16). Early surgery thus provides optimal assurance of a normal grip development. In addition, optimal normal development of the skeleton and muscles is ensured.

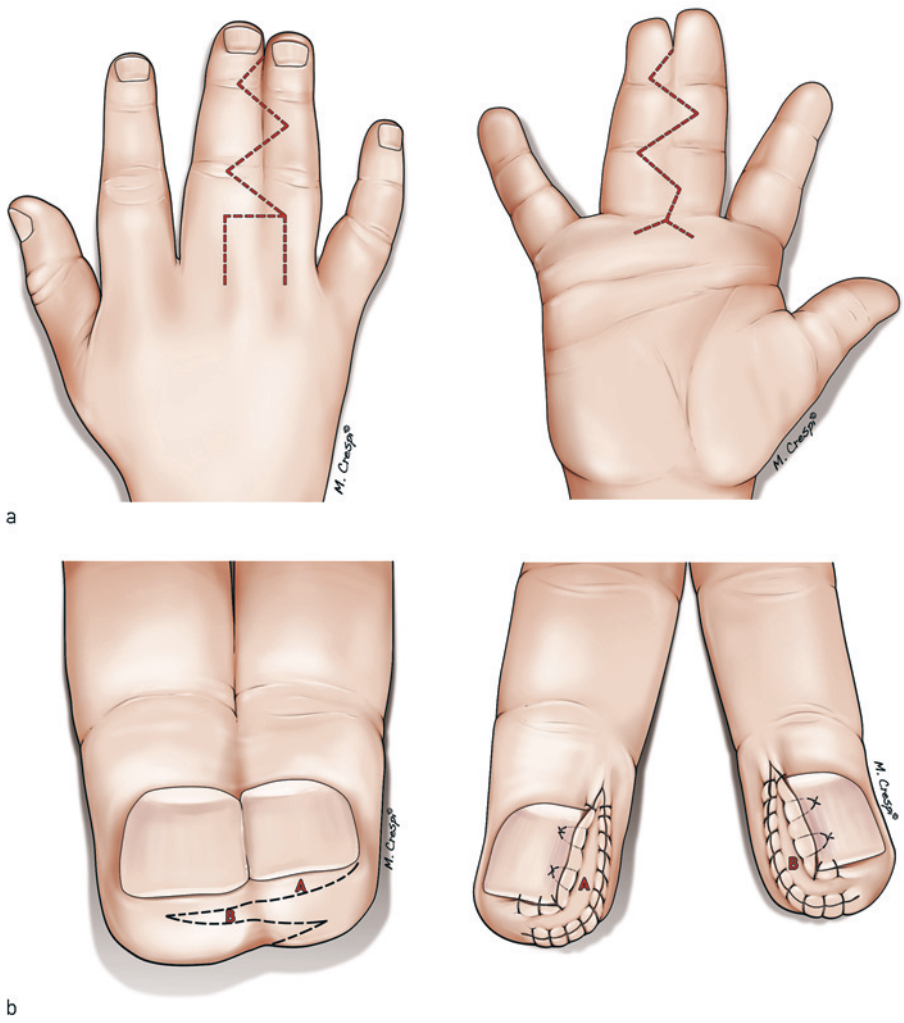

Figure 3 *Example of a plasty for separation of syndactyly between the long and ring finger. a) Planning sketch from the volar and dorsal aspect, where a dorsal rectangular flap for the web spaces and transposition flaps extending to the fingers are sketched. b) Planning sketch of distal phalanges with transverse pulp flaps and drawing in which these are sutured in place*

Information to the parents

It is important to establish a relationship of trust with the parents. This requires time and qualified answers to their questions and concerns about what can be achieved through surgery, as well as an exact account of the entire course of treatment. It is important to highlight the fact that it is not always possible to create a normal-looking hand, and that the main purpose of surgery is to raise the child's functional

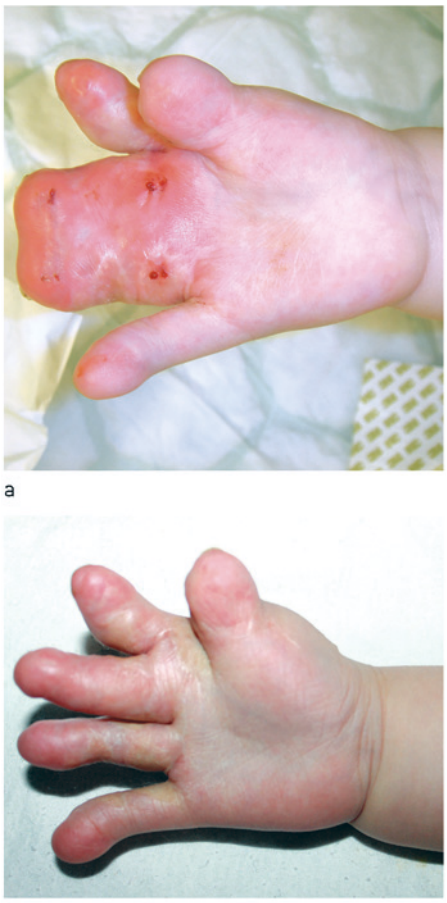

Figure 4 *Tissue expansion using external distraction device («magic cube») in a patient with complex syndactyly. Photograph of volar aspect of right hand in a patient with Apert syndrome who has had separation of complex syndactyly between all the fingers except between long and ring finger. a) Photograph following removal of «magic cube». b) Photograph of volar aspect after approximately one year following separation of long and ring finger*

level. The parents should be informed that several operations may be necessary to separate several fused fingers. The parents may have problems in understanding that the operation is not a simple separation of two fingers, but a reconstructive procedure which also involves the need for skin grafting. Children should be monitored until they have ceased to grow so that the development of web creep and scar contracture extending to the finger can be corrected before the child develops deformities.

Several of our university clinics have cross-disciplinary «dysmelia teams» consisting of a hand surgeon, an occupational therapist and an orthopaedic engineer to deal with the more complex conditions. The teams take care to provide information, and moreover parents and children can meet other children with the same or similar conditions, as well as their parents. In this way the coping aspect is also safeguarded.

Surgery

Each stage of the surgical procedure is designed to minimise the risk of complications. Surgery is performed under general anaesthetic with bloodless field and use of magnifying loupes, and microsurgical instruments must be available. If the condition is bilateral, the procedure is often carried out on both hands simultaneously. The various techniques for separation of fused fingers are largely based on the same principles. Both sides of a finger should not be operated in one procedure, as this can compromise circulation (1, 2, 12). An example of separation of a syndactyly is shown in Figure 3a. The bloodless field is released after the flaps are raised, and haemostasis is achieved before the flaps are inserted into place with absorbable sutures. Skin defects are covered with full-thickness skin grafts, often harvested from the groin. Use of split-thickness skin grafts is avoided, since these can shrink up to 50 % and thereby entail scar contractures (13). In syndactyly extending to the distal phalanges, the lateral nailfold is reconstructed with transverse fingerpulp flaps (Figure 3b) (14, 17).

If all the fingers are fused, the thumb and index finger, and the long and ring finger are separated during the first operation. During the second operation, normally 4–6 months later, the index finger and long finger, and the ring finger and small finger are separated (12, 15).

In case of syndactyly in children with Apert syndrome, it may be beneficial to cover the first web space with a rotational flap from the dorsal hand (1, 14, 15). With extensive syndactylies of this type, tissue distraction may be performed using an external distraction device (Cube fix distractor, developed by Rolf Habenicht at the Catholic Children's Hospital Wilhelmstift in Hamburg) before separating the last two fingers (long and ring fingers) (Figure 4) (18, 19). The device is referred to as the «magic cube» at Oslo University Hospital, since the distraction results in «extra» skin that makes subsequent separation easier.

Results of treatment

It is difficult to provide accurate data on treatment results, due to the heterogeneity of the condition, the many surgical techniques and the few long-term results published.

In simple syndactylies a good functional and cosmetic result is achieved (12, 20–22). However, a need for secondary procedures is reported in approximately 10 % of cases (12). Development of web creep occurred in two of 26 interdigital web spaces with the full-skin transplant technique after an 18-year follow-up period. (21). For the skin graft-free technique, long-term results after more than ten years' follow-up show a somewhat higher occurrence of the development of web creep (22).

Goldfarb et al. (23) demonstrated good range of motion following the separation of complex syndactyly between the long and ring finger or between the ring and small finger. However, rotational and axial deviation of the fingers, and nail deformity with inadequate lateral nail-folds were found on most of the fingers. The study is supported by Vekris et al. (24), who found that complex and complicated syndactylies, together with delayed separation, give poorer results. In a study of Apert syndrome without use of distraction, acceptable functional results were found (25). All the patients required corrective procedures, but taking into account the complexity of the condition, the need for revisions was considered acceptable.

There are good results from using the «magic cube» to separate the last two fingers (18, 19). The creation of a «five-fingered» hand was achieved, with the long and ring fingers having an appearance corresponding to the other separated fingers. Of eight syndactyly operations (19) there were two minor infections which were treated with antibiotics, and one infection following completion of distraction, in which the distraction device had to be removed. In this instance the surgery for syndactyly was postponed for three weeks. In one case the separated synostosis fused together again during distraction and had to be reoperated.

The most frequent postoperative complications related to syndactyly surgery are infection and maceration of the graft or flap which can result in their loss. Loss of fingers is rare, but is described in the literature (12).

Final comments

We present an overview of important principles in the care of children with syndactyly, where the main indication for surgery is to improve the functional level of the hand. Despite the fact that syndactyly is one of the most common congenital malformations of the upper extremities, the knowledge base is limited, since the condition is heterogenous and affects few patients. We present treatment results based on simple case series without randomisation. The studies conclude that safe separation of the fingers can be expected, and that these will function well independently. The treatment result is optimal for the least extensive conditions. For children with associated conditions and anomalies, it is favourable that the place of treatment should have all the necessary specialists. It is important that the parents are thoroughly informed. The parents' understanding of the course of treatment, and acceptance of what results can be expected,

are necessary to enable the child to cope with the condition.

Hebe Désirée Kvernmo (born 1961)
is a specialist in hand and orthopaedic surgery, Dr. med. and MHA. She is a senior consultant and President of the Norwegian Society for Surgery of the Hand.
The author has completed the ICMJE form and declares no conflicts of interest.

Jan-Ragnar Haugstvedt (born 1954)
is a specialist in general and orthopaedic surgery, and hand surgery. He completed his PhD after a research residency at the Mayo Clinic. He is a senior consultant and board member of the Norwegian Society for Surgery of the Hand, and deputy secretary general of the European Wrist Arthroscopy Society.
The author has completed the ICMJE form and declares no conflicts of interest.

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