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Features of surgical correction of various forms of hand syndactyly in children. Retrospective study of own treatment experience

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Syndactyly is a congenital malformation which is characterized by impaired differentiation of upper extremity tissues. Surgical correction of syndactyly is aimed to achieve satisfactory cosmetic and functional result. Most often, elimination of the total syndactyly form of the fingers implies is achieved by techniques according to Flatt (1962), Cronin (1943), Gilbert (1986), Wood (1998), bone form requires usage of Buck-Gramko technique. Objective. To conduct a retrospective study of surgical treatment results in patients with various forms of hand syndactyly. Methods. The study included 84 patients (109 hands) with hand syndactyly who were operated during the period from 2012 to 2020 in the pediatric orthopedics clinic of the Sytenko Institute of Spine and Joint Pathology National Academy of Medical Sciences of Ukraine. The mean age of patients was 6.5 years (1 to 16), 39 (46.4 %) boys and 45 (53.6 %) girls. Most often syndactyly of III–IV fingers (105 (96.3 %) hands) was managed by the Wood method, namely in 63 (60.0 %) hands and 8 (7.6 %) cases with severe bone forms were corrected by Buck-Gramko method. Rotational skin pieces Ghani and Buck-Gramko were used for surgical correction of I–II fingers syndactyly. Treatment results were evaluated by the Vancouver Scar Scale (VSS). Results. According to VSS, the treatment result was classified as satisfactory in 73 (67.0 %) hands. Complications were noted in 11 (10.1 %) cases: 2 patients (18.2 % of 11) with congenital amniotic membranes were found to have lysis of a free skin piece; 1 (9.1 %) after removal of the bony syndactyly form had deviation of the nail phalanx; 3 (27.3 %) with Poland-syndrome were shown to have scarring of the interdigital space; 5 (45.4 %) with a complex bony form of syndactyly further on developed pulling scars, which caused deformity of the fingers and resulted in a correction in the form of multistage Z-plastics. Conclusions. All the patients showed improvement in the function and cosmetic results of the hand at the end of treatment. The best results were obtained in the case of simple and total forms of syndactyly treated with Wood technique. Key words. Children, congenital anomalies of the hand, syndactyly, surgical correction.

Синдактилія — вроджена вада розвитку, що характеризується порушенням диференціації тканин верхньої кінцівки. Хірургічна корекція синдактилії спрямована на досягнення задовільного косметичного та функціонального стану кистей. Найчастіше для усунення тотальної форми синдактилії пальців застосовують методики за Flatt (1962), Cronin (1943), Gilbert (1986), Wood (1998), за умов кісткової форми виконують методику Buck-Gramko. Мета. Провести ретроспективний аналіз хірургічного лікування пацієнтів із різними формами синдактилії кисті. Методи. За період із 2012 по 2020 рік у клініці дитячої ортопедії ДУ «ІПХС ім. проф. М. І. Ситенка НАМН України» прооперовано 84 пацієнти (109 кистей) із синдактилією кистей. Середній вік хворих становив 6,5 року (від 1 до 16), 39 (46,4 %) хлопчиків і 45 (53,6 %) дівчинок. Найчастіше для релізу синдактилії III–IV пальців (105 (96,3 %) кистей) використано методику за Wood — 63 (60,0 %) кисті, за умов важких кісткових форм у 8 (7,6 %) випадках — методику Buck-Gramko. Для хірургічної корекції синдактилії I–II пальців застосовано ротаційні шкірні шматки Ghani та Buck-Gramko. Результати лікування оцінювали за Vancuver Scar Scale (VSS). Результати. За VSS результат лікування класифіковано як задовільний на 73 (67,0 %) кистях. На 11 (10,1 %) відмічені ускладнення: у 2 хворих (18,2 % від 11) із вродженими амніотичними перетинками — лізис вільного шкірного шматка; у 1 (9,1 %) після усунення кісткової форми синдактилії — девіацію нігтьової фаланги; у 3 (27,3 %) з Poland-синдромом — рубцювання міжпальцевого проміжку; у 5 (45,4 %) зі складною кістковою формою синдактилії в подальшому формувалися тягнучі рубці, що спричиняло деформацію пальців, та обумовило корекцію у вигляді багатоступеневої Z-пластики. Висновки. У всіх пацієнтів спостерігали покращення функції та косметичного вигляду кисті наприкінці лікування. Кращі результати отримано в разі хірургічного лікування простих і тотальних форм синдактилії за методикою Wood.

Key words. Children, congenital anomalies of the hand, syndactyly, surgical correction

Introduction

Despite the improvement of prenatal and intranatal diagnosis of fetal developmental abnormalities, congenital anomalies of the hand today are an important part of orthopedic pathologies. A significant number of them are diagnosed as syndactyly, an abnormality characterized by impaired differentiation of limb tissues [1–3].

The incidence of syndactyly in the general population is 1 in 2,000–2,500 newborns, 50 % of whom have bilateral pathology [4–7]. Syndactyly in 10–40 % of cases is an isolated autosomal dominant hereditary disease. Syndromic manifestations of this disease are associated with many genetic disorders, such as: Poland, Apert, Holt-Oram, Carpenter syndromes and congenital amniotic membranes [1–5]. In most cases, I and II, II and III finger syndactyly is diagnosed in conditions of genetic syndrome [6]. In contrast to genetically determined one, the isolated form is characterized by the fusion of III-IV fingers.

Syndactyly is classified according to the following features: simple type as the fusion of only the skin of the fingers and severe with bone fusion and deformation of the fingers. There are basal (skin fusion to the level of the middle phalanx of the fingers) and total (fusion of fingers along the entire length) forms. The classification of S. A. Temtamy and V. A. McKusick [8] is widely used.

Surgical correction is indicated in all cases of syndactyly, as it is aimed at improving the function of the hand and cosmetic appearance [8]. Contraindications for intervention may be incomplete syndactyly, which does not affect the function of the hand, or a total severe form, when the separation of the fingers does not result in improved function [9, 10]. The purpose of the operation is to create an adequate, sufficiently deep gap between the fingers and improve their cosmetic appearance.

The classic method of releasing syndactyly using Z-shaped approaches according to Bruner is generally accepted, which provides partial coverage of the lateral surfaces of the fingers with local tissues, with the obligatory formation of the interdigital space at the expense of own tissues. All known methods of syndactyly release differ from each other only in the type of skin flap during the formation of the interdigital space. Most often, Flatt (1962), Wood (1998), Cronin (1943), and Gilbert (1986) methods are used to eliminate the total form of finger syndactyly, and Buck-Gramko (Fig. 1) in the case of the bony form. Most surgeons try to cover the lateral surfaces of the fingers only with local triangular pieces of skin, but the literature describes a sufficient number of complications in the form of pulling postoperative scars [11, 12].

That is why, usually, combined skin plastics with loose skin pieces are used [13–15]. In cases of genetically determined syndactyly of the I – II fingers, different types of posterior rotational skin pieces are used, in particular according to the Ghani and Buck-Gramko method (fig. 1, d). This makes it possible to create a wide and deep first interdigital space, which is the main one for the optimal function of bilateral hand grip [16–19].

A method of soft tissue distraction using the «magic cube» / «cube fix» is also described, which is administered in the case of total and complex forms of syndactyly or pansyndactyly with bone fusion secondary to Apert syndrome [20–23]. The method was proposed by Dr. Habenicht. Distraction allows the soft tissues to be stretched to increase their area, which further improves the formation of triangular pieces and the covering of the lateral surfaces of the fingers with local tissues. Due to technical features, soft tissue distraction can be achieved only at the level of the distal and middle phalanges of the fingers, and after the formation of the interdigital space there are skin defects that need to be covered with free pieces of skin [24, 25].

The issue of the age when children with syndactyly should be treated remains to be open. Some authors consider it appropriate to perform operations from 6 months, but most of the world's experts in pediatric hand surgery recommend doing it from 12 months [23, 26, 27]. In severe bone forms, it is recommended to start treatment at 12 months. Pansyndactyly or syndactyly secondary to a genetic syndrome with lesions of I and II, or IV and V fingers needs to be eliminated as a matter of priority, taking into account the different length of the fingers and the possible development of deformities with rapid growth of the child.

The aim of the study: to analyze the results of surgical treatment of patients with syndactyly, operated at the pediatric orthopedics clinic of the Sytenko Institute of Spine and Joint Pathology National Academy of Medical Sciences of Ukraine.

Material and methods

The materials of the article were considered at the meeting of the Committee on Bioethics at the Sytenko Institute of Spine and Joint Pathology National Academy of Medical Sciences of Ukraine and received a positive decision (Minutes No. 216 as of 26.04.2021).

In the period from 2012 to 2020, 84 patients (109 hands) with congenital anomalies in the development of the upper extremities were operated on. The average age of patients was 6.5 years (1 to 16), 39 boys (46.4 %) and 45 girls (53.6 %). Unilateral syndactyly was diagnosed in 59 (70.2 %), of which right-sided in 35 (59.3% of 59 patients), left-sided in 24 (40.7 %). Three forms of syndactyly were found in children with unilateral pathology, namely basal in 19 (32.2 %), total in 28 (47.5 %), total with bone fusion in 12 (20.3 %).

Bilateral syndactyly was detected in 25 (28.1 %) children. Among them, 13 (52.0 %) children were found to have genetic abnormalities: 3 (23.1 % of 13 patients) had Apert syndrome, 8 (61.5 %) Poland syndrome, 2 (15.4 %) Holt-Oram syndrome. Congenital amniotic membranes were diagnosed in 12 (48.0 %) patients.

All patients were pre-examined, if necessary, underwent radiography. Surgical treatment was performed according to functional and cosmetic indications. In the case of pansyndactyly, syndactyly of the I–II and IV–V fingers were first separated.

The most frequently employed methods to eliminate syndactyly of the III–IV fingers (105 (96.3 %) hands) in our practice were as follows: by Wood in 63 (60.0 %) cases (fig. 2), by Gilbert in 21 (20.0 %), by Cronin in 13 (12.4 %). They were performed according to the basic classical principles of surgical approaches using both local tissues and loose skin pieces. Under conditions of severe bony forms of syndactyly in 8 (7.6 %) cases the Buck-Gramko method was used.

Various variations of rotational dorsal vascularized skin pieces were used to correct the syndactyly of the I–II fingers (4 (3.7 %) of the hand), the most common being Ghani, which allowed to form a sufficient width and depth of the interdigital space for the correct position of the I finger.

Postoperative results were evaluated using the Vancouver Scar Scale (VSS) [28, 29]. It is used to qualitatively assess the cosmetic and functional condition of the operated hand in terms of pigmentation, vascularization, mobility of skin pieces and the height of the postoperative scar (table). The main qualitative indicators were the height of the postoperative scar and the mobility of the skin piece. The index of the postoperative scar more than 2–4 mm indicated the presence of its hypertrophy, which can result in contractures of the fingers of the hand and indicate unsatisfactory cosmetic and functional results of surgery.

Results and their discussion

According to the VSS, 109 operated hands were evaluated and the result was classified as satisfactory in 73 (67.0 %) of them. At the same time good vascularization of a skin piece, sufficient mobility

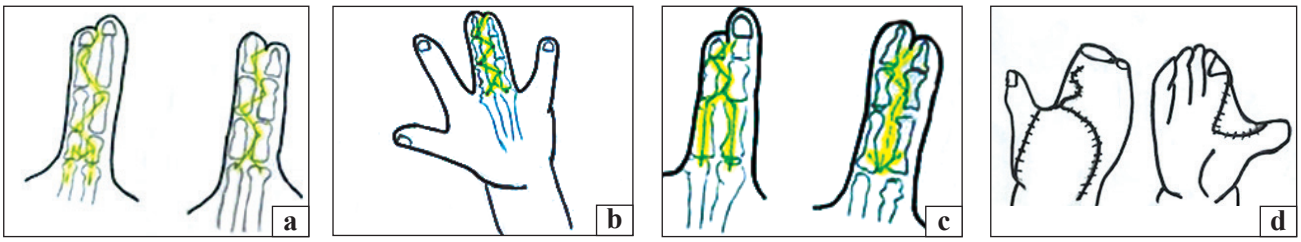

Fig. 1. Schematic representation of surgical approaches: a) according to Wood; b) Cronin; c) Gilbert; d) Ghani dorsal flap

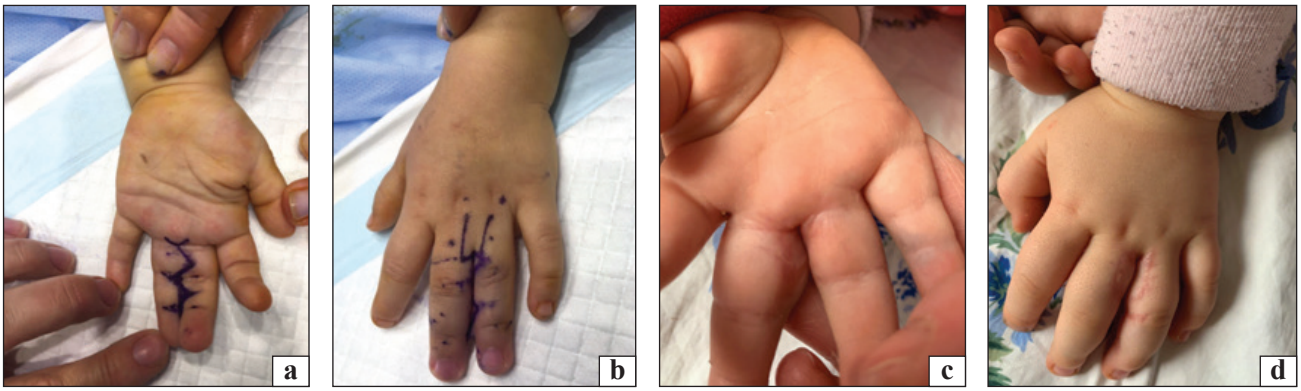

Fig. 2. Appearance of a 1-yer-old patient's Ya. hand: intraoperative marking according to Wood (a, b); in 2 months after surgery (c, d)

Table

Evaluation of the postoperative area by Vancouver Scar Scale (points)

Score	Pigmentation	Vascularization	Mobility	Scar height
0	normal	normal	normal	normal
1	hypo-pigmentation	pink integuments	elastic	> 0–1 mm
2	mixed	red integuments	pliable	> 1–2 mm
3	hyper-pigmentation	crimson integuments	firm	> 2–4 mm
4	—	—	contracting	> 4 mm
5	—	—	contracture	—

of a recipient zone of skin pieces and a site of local plasticity was established. The height of the postoperative scar did not exceed 1 mm, which gives a possibility to assess the postoperative result as satisfactory. However, complications were noted on 11 (10.1 %) operated hands: 2 patients (18.2 % of 11) with congenital amniotic membranes were found to have lysis of the free skin piece, caused by infection due to intradermal fistulas; 1 (9.1 %) after removal of the bony form of syndactyly was shown to have deviation of the nail phalanx; 3 (27.3 %) with Poland-syndrome had scarring of the interdigital space, associated with the early age of the child and severe hypoplasia of the fingers with a sharp reduction in the number of local tissues; 5 (45.4 %) with a complex bony form of syndactyly subsequently formed pulling scars, which caused deformity of the fingers and required correction in the form of multistage Z-plasty.

Despite the complications developing after surgical treatment of complex forms of syndactyly, all patients had an improvement in the function and cosmetic appearance of the hand.

Conclusions

The most difficult aspects in the treatment of syndactyly:

– Poland-syndrome with severe hypoplasia of the fingers and a sharp reduction in the number of local tissues, accompanied by scarring of the interdigital space;

– secondary to congenital membranes with the formation of intradermal fistulas, which can be complicated by lysis of the skin piece in case of infection;

– bony, total forms with further development of deviation of the nail phalanx and formation of immobile postoperative scars with limited movement in the interphalangeal joints of the fingers.

The best results were obtained in the case of surgical treatment of simple and total forms of syndactyly according to the method of Wood. In our opinion, all patients with syndactyly should perform surgery in preschool age not only for functional and cosmetic purposes, but, above all, for good social adaptation of the child.

Conflict of interest. The authors declare no conflict of interest.

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FEATURES OF SURGICAL CORRECTION OF VARIOUS FORMS OF HAND SYNDACTYLY IN CHILDREN. RETROSPECTIVE STUDY OF OWN TREATMENT EXPERIENCE

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