

Original Paper

Particularities and Surgical Treatment of Constriction Band Syndrome Syndactyly, in Children

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REZUMAT

Particularitățile și tratamentul chirurgical al sindactiliei secundare maladiei amniotice, la copii

Introducere: Sindactilia secundară maladiei amniotice este o afecțiune sporadică, prezentă deseori în asocieri cu alte patologii precum malformații musculo-scheletale, cranio-faciale, toraco-abdominale, defecte de tub neural sau hemangioame. S-a constatat o afectare mai frecventă a membrilor superioare (măinile sunt afectate în cca. 90% din cazuri), degetele mai lungi fiind mai des implicate decât cele scurte. Dintre malformațiile secundare maladiei amniotice prezente la nivelul mâinilor fac parte sindactilia, acrosindactilia, hipoplazia digitală, campodactilia, amputația și limfedemul.

Material și metode: În studiu au fost incluși 33 de pacienți (cu un total de 98 de spații interdigitale afectate) operați pentru sindactilia secundară maladiei amniotice în perioada 2005-2015 pe secția de Chirurgie Plastică a Spitalului Clinic de Urgență pentru Copii "Grigore Alexandrescu". La acești pacienți au fost urmărite și notate: vârsta pacienților în momentul începerii tratamentului chirurgical, numărul și durata intervențiilor chirurgicale, necesitatea grefelor tegumentare, prezența complicațiilor intraoperatorii și postoperatorii, perioada de urmărire postoperatorie, complexitatea malformațiilor (prezența hipoplaziei digitale, amputațiilor și limfedemului).

Rezultate: Șase dintre pacienții incluși în studiu au beneficiat de câte două intervenții chirurgicale fiecare, optsprezece au necesitat între trei și șapte operații și nouă dintre ei au avut cel puțin opt intervenții chirurgicale (între opt și doisprezece operații). Pentru cincizeci și trei dintre spațiile interdigitale operate au fost necesare grefe tegumentare, pe când pentru reconstrucția a patruzeci și cinci de spații nu au fost utilizate grefe. La nici unul dintre cei 33 de pacienți operați nu s-a constatat postoperator liza grefelor tegumentare, necroza lambourilor sau apariția infecției.

Concluzii: Maladia amniotică cuprinde o asocieri complexă de malformații asimetrice, fiecare caz fiind unic din punctul de vedere al manifestărilor clinice. Prognosticul sindactiliei secundare acestui sindrom depinde de severitatea anomaliilor asociate. Tratamentul chirurgical precoce este necesar în vederea obținerii unor rezultate satisfăcătoare.

Cuvinte cheie: acrosindactilie, maladie amniotică, inel de constricție, tablou clinic heterogen, tratament chirurgical

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ABSTRACT

Introduction: Constriction band syndrom syndactyly is a sporadic condition, that may be present in association with other congenital anomalies such as musculoskeletal, craniofacial and thoraco-abdominal disorders, neural tube defects, scoliosis and hemangiomas. There is a significant predilection for the upper extremities and an increased frequency in distal limbs, and longer digits are significantly more involved than shorter ones. The hands are affected in almost 90% of cases. Associated hand anomalies include syndactyly, acrosyndactyly, digital hypoplasia, symbrachydactyly, campodactyly and lymphedema.

Materials and methods: We reviewed 98 webs in 39 hands and 7 feet in 33 patients, operated for congenital constriction band syndactyly during the period of 2005 -2015 in the Plastic Surgery and Burns Department of "Grigore Alexandrescu" Clinical Emergency Hospital for Children. Number and duration of surgical procedures, age starting surgery, the need for skin graft ,intraoperative and postoperative complications (development of web creep, necrosis, flection contracture, finger deviation), follow up period, malformation complexity (digital hypoplasia, amputation, lymphedema) were assessed clinically and documented.

Results: Six of the patients included in the study received two surgeries each, eighteen needed between three and seven surgeries and nine of them required at least eight surgeries (between eight and twelve operations). Fifty-three of the reconstructed webs needed skin grafts to cover the aria resulting from release of digits, and forty-five didn't need skin grafts. Of the 33 operated patients, there were no cases of skin graft loose or flap necroses, and none of the patients developed local infection due the surgery.

Conclusions: The constriction ring syndrome is a complex collection of asymmetric congenital anomalies, in which no two cases are exactly alike. The prognosis is dependent on the severity of the associated defects. Early intervention is needed for a successful outcome.

Key words: acrosyndactyly, constriction band syndrome, constriction ring, heterogeneous expression, surgical treatment

INTRODUCTION

Constriction band syndrom syndactyly is a sporadic condition, that may be present in association with other congenital anomalies such as musculo-skeletal (including total amputation of the limb), craniofacial (like cleft lip and cleft palate) and thoraco-abdominal disorders (such as gastroschisis and extrathoracic heart), neural tube defects, scoliosis and hemangiomas (1, 2, 3, 4, 5, 6). Some of these manifestations are uncommonly noted at birth because they result in spontaneous abortion.

There are two main theories about constriction band syndrome etiology, the first of which is the intrinsic theory proposed by George Streeter, the director of embryology at Carnegie Institute, in 1930 which is called, "Streeter's Dysplasia". Streeter suggested that an intrinsic defect in the subcutaneous germplasm resulted in focal mesenchymal hypoplasia, tissue loss, and scarring (7, 8). Another theory, the extrinsic theory was proposed in 1965 by the obstetrician Richard Torpin and is known as the Amniotic Band Syndrome (ABS). Proceeding rupture, the amniotic sac stops growing properly and eventually

separates from the chorion. The chorionic side of the amnion emanates numerous mesoblastic fibrous strings which entrap and catch the fetal parts. This then leads to the constriction of the body part stopping blood flow to that area, leading to fetal amputations as one of many consequences. (9)

The incidence of constriction band syndrome is approximately one in 1200 to one in 15000 live births, and affects both sexes with a ratio of 1:1 (1, 2, 6, 10).

There is a significant predilection for the upper extremities, an increased frequency in distal limbs and longer digits are significantly more involved than shorter ones. The hands are affected in almost 90% of cases. Associated hand anomalies include syndactyly, acrosyndactyly, digital hypoplasia, symbrachydactyly, camptodactyly and lymphedema (2, 6).

Patient with untreated constriction ring syndrom malformations ,affecting all limbs (Fig. 1,2,3,4,5).

The main treatment of the anomalies secondary to congenital constriction band syndrome is by surgery. Timing , order and technique of repair are dictated by disease severity and predicted skeletal growth. Because of the unique presentation of deformations in each individual, they represent a



Figure 1. Patient with untreated constriction ring syndrom malformations, affecting all limbs. Right hand with two digits amputated - dorsal view



Figure 2. Patient with untreated constriction ring syndrom malformations, affecting all limbs. Right hand with two digits amputated - volare view



Figure 3. Patient with untreated constriction ring syndrom malformations, affecting all limbs. Right upper limb amputation at the elbow



Figure 4. Patient with untreated constriction ring syndrom malformations, affecting all limbs. Right foot with great toes amputation

treatment challenge (11).

The aim of the present study was to evaluate the results from surgical correction and particularities of congenital constriction band syndactyly in children, treated in the Plastic Surgery and Burns Department of “Grigore Alexandrescu” Clinical Emergency Hospital for Children between 2005 and 2015.

MATERIALS AND METHODS

We reviewed 98 webs in 39 hands and 7 feet in 33 patients, operated for congenital constriction band syndactyly during the period of 2005 -2015 in the Plastic Surgery and Burns Department of “Grigore Alexandrescu” Clinical Emergency Hospital for Children. All the webs have been corrected by the same surgeon, using the same indication, timing and technique.

None of the patients describe positive family history of amniotic band syndrome and twelve of the



Figure 5. Patient with untreated constriction ring syndrom malformations, affecting all limbs. Left foot with first four toes syndactyly and tibial deviation

patients associated congenital disorders (upper limb amputation, cleft lip, cleft palate, hemangiomas, talipes equinovarus). (Fig. 6,7,8)

More than half of patients (55 %) included in the study were born prematurely.

Age at the time of first surgery varied from four months to seventy months. Seventeen of the patients were boys and sixteen girls.



Figure 6. Constriction rings above the ankle of the both legs. Right foot talipes equinovarus



Figure 7. Severe banding of both hands resulting in multiple digits amputation and acrosyndactyly

Of these patients, twenty-seven of them had only one superior limb affected and six of them had both upper limbs and lower limbs affected. Seven hands had only one web involved, eight had two webs affected and eighteen had more than two webs with syndactyly. Foot syndactyly affected twenty-one webs.

Number and duration of surgical procedures, age starting of surgery, the need for skin graft, intra-operative and postoperative complications (development of web creep, necrosis, flexion contracture, finger deviation), follow up period, malformation complexity (digital hypoplasia, amputation, lymphedema) were assessed clinically and documented. Web-creep was objectively and serially assessed using the volar base of the web as a baseline. Scar quality was assessed using Vancouver Scar Scale (12).

Follow-up period ranged from 17 to 160 months with an average of 72 months.

Surgical consideration

The surgery is done under general anesthesia and tourniquet control.

Surgical approach is guided by the severity of the disease. Acrosyndactyly treatment in the first 6 months allows the best chance for proper longitudinal bone growth. This is particularly important when the joined fingers are of different lengths.

For the commissural reconstruction of the first web there have been used the four - flaps Z-plasty technique, while the rest of the webs have been released using the technique of Flatt modified by Upton (1990). Beyond the flaps used in commissure reconstruction, the fingers are separated using zigzag incisions. The palmar flaps are based opposite the dorsal flaps (mirror images) to allow interdigitation. This orientation minimizes the tendency for



Figure 8. Last three fingers left hand acrosyndactyly in amniotic disruption sequence

the formation of a flexion scar contracture development and maximizes coverage potential (13). The separation of the digits requires the division or excision of the fascial interconnections between the digits, with care taken to identify and preserve the individual neurovascular bundles, as well as to ensure adequate venous drainage of commissure (14, 15). The bare areas on both lateral sides are covered with skin grafts. All the skin grafts were full - thickness and they were harvested

Table 1. Peculiarities of the studied group

Particularities	Numbers
Patients/ webs	33/98
Male / female	17/16
Age at first syndactyly release surgery (months)	4 – 70
Hands/ foot syndactyly (webs)	77/21
Patients with /without other congenital anomalies	12 / 21
Simple/ complicated syndactyly (webs)	31/ 67
Follow-up period (months)	17- 160
Preterm / term infants	18/ 15

from the distal wrist crease or from the groin area.

Cosmetic repair of shallow constriction rings that present without distal edema may be done electively. Release of bands associated with severe distal edema should be performed within the first few days after birth. For release of superficial and deep onstriction rings, in the presence of good distal function, Z- plasty, W- plasty and V-Y flaps are the mainstay. Deep rings with neurovascular compromise may require special attention to neurovascular reconstruction.

The postoperative dressing must apply compression across the skin grafts and protect the separated digits, but care must be taken not to apply strong compression. The fingertip is kept visible at all times to check the vascularity (16). The compression dressing is removed at one week after surgery and a light dressing is applied until two weeks postoperatively.

For the adjacent side a delay of 6 months is proposed before separating the second side. Usually the patients are discharged after two days.

After reconstruction, patients should be examined periodically since these patients require multiple surgeries until they have achieved skeletal maturity.

RESULTS

Six of the patients included in the study received two surgeries each, eighteen needed between three and seven surgeries and nine of them required at least eight surgeries (between eight and twelve operations). The last group consists of patients with severe acrosyndactyly and bilateral upper limb impairment. (Fig. 9,10,11)

Twenty-seven of the patients' surgeries started in the first year of life while six of the patients had the first surgery after the age of one year. Patients who came to the surgeon after the age of one year showed mild acrosyndactyly and the majority (five of them) were from rural areas.

Fifty-three of the reconstructed webs needed skin grafts to cover the area resulting from release of digits, and forty-five didn't need skin grafts. Hair growth has been found on four webs and hyperpigmentation in five webs, all grafted with skin from the groin. None of the webs grafted with skin from the distal wrist crease developed hyperpigmentation or hair growth. Of the 33 operated patients, there were no cases of skin graft lose or flap necroses, and none of the patients developed local infection due the surgery.



Figure 9. Bilateral hand constriction band syndrom - after surgery outcome, dorsal view



Figure 10. Bilateral hand constriction band syndrom, after surgery outcome - volare view



Figure 11. Amniotic band syndrome amputation defect of digits of both legs - after two surgeries

As to the duration of surgical procedure, there was a range from 30 to 100 minutes, with a general average of 50 minutes. There was a difference in duration of surgery between simple cases (38 minutes) and complicated cases (75 minutes). It was found that the average length and number of surgeries, the rate of complications and relapses are higher compared to primary syndactyly treatment. This is due to malformations complexity which increase the difficulty and number of surgeries.

Web creep was seen in 28 of the 78 webs operated before 1 year of age, while from the 20 webs operated later, only 3 developed web creep. The most of the patients treated in the first year of life had

complex or complicated syndactyly, associated with digital hypoplasia, symbrachydactyly, camptodactyly, constriction rings, lymphedema, interphalangeal joints disorders or finger amputation, which means that the difference in the incidence of late complications correlate not only with the timing of surgery but also with the complexity of this congenital anomaly. Twenty-eight of the web creep required surgery. The revisional surgery was performed using the same technique and in seven of the reinterventions skin graft was required.

The scar quality evaluation revealed a height below 2 mm in 93 of the 98 spaces, normal or supple pliability in 91 of the webs. There was no evidence of flexion contracture. All patients had good finger-tip sensation.

DISCUSSION

Amniotic band syndrome is a set of multiple malformation due the adhesion and indentation caused by amniotic bands during gestation (6).

Prenatally, amniotic band syndrome may be detected by ultrasonography with the observation of asymmetrical limb deformities or defects and visualisation of the amniotic membranes wrapping around foetal portions (16, 17, 18). The indications for fetal surgery in the amniotic band syndrome may be either for a life-threatening condition, if it involves constriction of the umbilical cord, or more commonly, threatened limb amputation due to amniotic band constriction. The rationale for performing fetoscopic lysis of constricting extremity amniotic bands is based on the hypothesis that progressive compromise of fetal growth leads to amputation. However, this assumes that the procedure can be accomplished with no maternal morbidity and minimal fetal morbidity. This procedure would be hard to justify in the face of a serious maternal complication or a fetal death due to severely premature delivery at 21 or 23 weeks of gestation, even in the face of certain fetal limb amputation. However, the sequelae of the ABS may not completely remove or may result in

secondary lymphedema.

Physical examination is the main way of postnatal diagnosis of constriction band syndrome, with usage of different investigations in order to establish potential malformations of different organs and body parts: X-ray, ultrasound (16).

In the hand, clinical presentation of amniotic band syndrome varies from slight indentations on the affected part to distal atrophy, lymphedema, acrosyndactyly, interphalangeal joints disorders and amputation. The treatment of acrosyndactyly differs depending on the extent of fusion and associated band deformities. Each case is unique and must be individualized.

Frequent association of hand deformities with other congenital disorders (19) (in our study 33,6% of the cases), some of them with the indication of treatment before the syndactyly release, is the reason why these patients require careful assessment and investigation at birth, and a multidisciplinary approach.

The goal of the constriction band acrosyndactyly treatment is to increase function of the hand, to foster the development of fingers while providing a more aesthetic appearance, using the fewest surgical procedures while minimizing complications. Many children with amniotic band syndrome require multiple surgeries, and the family and surgeon often work together for quite some time.

In general, the fingers are separated using carefully planned Z-plasty skin flaps, and a broad commissural space is created based on local skin flaps. The sinus tract is excised and may be used as a skin graft. This tract is generally not well suited for a commissural space because of its distal location and narrow space. Skin flaps are mobilized, and the tissue is defatted (17). Fingers are separated proximally to the transverse intermetacarpal ligament. Blood vessels and nerves are preserved, and the skin flaps are sewn down, with a full-thickness skin graft covering the bare area (20). In order to achieve good results from syndactyly release it is not enough that the main surgeon to be well acquainted with the anatomy of the hand. To minimize the rate of complications, this type of surgery should be performed in a plastic surgery department of a hospital for children, by a surgical team experienced in treating the child syndactyly, using appropriate surgical instruments and loupe magnification. (21) When graft failure occurs, the area should be regrafted to avoid scars because these tend to tether the joint as

Table 2. Need for skin graf

Type of syndactyly	Number of webs released using skin graft	Number of webs released without skin graft
Simple	16	15
Complicated	37	30
Web creep	7	21

the patient grows.

Because phalangeal amputation often occurs through the diaphysis, diaphyseal overgrowth is a recurrent problem in early amnion rupture sequence hands prior to the skeletal maturity. It is therefore important to avoid extensive distal skin grafting to retain pointed digital tips (13).

When band indentation is mild without distal edema, the appearance may be improved by excision of the indented skin band and Z plasty skin flap rearrangement, V-Y plasty or W plasty. All ring tissue is considered scar and it should be removed. When band indentation is accompanied by distal edema, excision of constriction band reduces lymphedema, and redundant skin excision and flap debulking may further improve soft-tissue contour (11).

Timing of the operation is an important matter of discussion in hands amniotic bands syndrome anomalies. In the patient with acrosyndactyly, finger release in the first 6 months allows the best chance for proper longitudinal bone growth. This is particularly important when the joined fingers are of different lengths. Cerebral pattern hand use after separation of digits is optimized when the web reconstruction is performed in the first year of life (22). Follow up should be until skeletal maturity, mainly due to the prevalence of web creep until this age (23).

CONCLUSIONS

The constriction ring syndrome is a complex collection of asymmetric congenital anomalies, in which no two cases are exactly alike. The prognosis is dependent on the severity of the associated defects. The syndrome and its complications can be easily diagnosed clinically and are amenable to corrective surgery with good results. Improvements in ultrasound imaging techniques and extension of the indications for fetoscopic examination could lead to an optimization of the prenatal diagnosis and treatment of amniotic bands syndrome by minimally invasive fetal surgery. Management of amniotic band syndrome syndactyly is focused on increasing function and development while providing an more aesthetic appearance. Early intervention is needed for a successful outcome. The parents should be informed from the start that the surgical treatment is a reconstructive procedure that may require multiple interventions.

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