

**CASE REPORT**

# Mutaf one-stage technique for congenital constriction rings – safe and effective

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**Summary**

**Introduction:** Congenital constriction ring is a rare congenital malformation that especially affects the limbs. It is associated with fibrous bands that could encircle, strangle, or amputate some parts of the fetus. However, there are only a few techniques described for the correction of this anomaly. We aim to present a successful treatment of congenital constriction ring by means of the one-stage Mutaf technique.

**Patient Review:** In our report, we presented two infants with congenital constriction rings. In the first case, we presented an infant with a constriction band on the right arm accompanied by deformity of the distal part and lymphoedema. In the second case, we presented an infant with multiple constriction bands on the distal part of the right leg with clubfeet and severe lymphoedema. They were both treated by the one-stage Mutaf technique using rectangular dermofat grafts to fill the groove. Using this technique we obtained a normal limb contour, with no oedema or neurovascular compromise. Filling the groove with the rectangular dermofat flaps parallel to the relaxed skin tension lines provides a better scar in comparison with multiple Z- or W-plastics techniques. Additionally, advantages of the one-stage procedure are reduction of surgical and psychological trauma in patients and their parents, less time spent under general anesthesia and lower amount of anesthetic the patient receives, and lower treatment costs.

**Conclusion:** We find the Mutaf technique to be an effective and safe technique for the treatment of congenital constriction rings.

**Keywords:** Amniotic Band, Surgery, Technique

## INTRODUCTION

Constriction ring syndrome (CRS) is a complex of congenital anomalies that occurs in a variety of combinations, especially affecting the limbs and rarely the head and the trunk. (1-10) There are several synonyms for this condition such as amniotic band syndrome (ABS), amniotic constriction bands (ACB), congenital constriction band syndrome (CCBS), annular band, and Streeter dysplasia. (7-18) This congenital malformation occurs with an incidence of approximately 1:1200 to 1:15000 live births. (2,7,14,18) It is characterized by fibrous bands -congenital constriction rings (CCR) - that could cause acrosyndactylies, circular constrictions in the extremities of varying degree, and terminal transverse defects. (1-3,5-7,9) There are several classification systems of this anomaly. (2,6,7) Patterson diagnostic criteria established a classification system of CCR: type 1 – simple constriction rings; type 2 – constriction rings accompanied by deformity of the distal part, with or without lymphoedema; type 3 – constriction rings accompanied by fusion of distal parts, ranging from mild to gross acrosyndactyly (type I, II, and III); and type 4 – intrauterine amputations. (2,4-7) Hall, Winzweig, Hüsler, Hannigan and Homer presented their classifications of CRS. (2,6,7)

The CRS is an independent group of congenital anomalies that occurs sporadically without a genetic background. (1,7) The etiology of this syndrome is still unknown. (1-3) There is no data about the inheritance pattern or chromosomal disorder and there is no difference in morbidity concerning gender and ethnicity. (1-9) The most common etiologies that have been discussed are the intrinsic theory (the umbilical cord causes pressure on the fetal tissue acting like constricting band and thus interfering with normal limb development) and the extrinsic theory (external maternal injury can cause an oligoamnion which can allow a closer contact between the fetus and the amnion). (3,4) However, no single pathogenic conclusion has been made to explain different findings seen in this syndrome. (1,3,7)

Soft tissue, lymphatic vessels and, superficial vascular circulation are usually partially obstructed and oedema with cyanosis could be distally present causing pain. (2,5,7-9) The prenatal ultrasound scan can show swelling of digits or limbs distal to the constriction with amniotic bands attached to the fetus and a reduction in fetal movements. (8)

There are several reports presenting the fetoscopic release of CCR. (20) However, in most cases the treatment of CCR occurs after birth. (1,5,9,11-19) When there are shallow grooves, surgery is not required. (1,2,11) In case of vascular or lymphatic obstruction, constriction bands must be surgically treated to prevent gangrene or auto-amputation and to save the function and development of the affected limb. (9-12,14,15,18) A surgical treatment is usually performed as a one-stage or two-stage procedure,

including multiple Z-plastics, W or Y-to-V plastics (with a variety of techniques proposed by different authors), direct closure, and rectangular-plastic technique (a one-stage Mutaf procedure). (1,9,11,12,14-19).

We present our results using the one-stage Mutaf technique in two patients with limb obstructions caused by CCR.

## Surgical technique

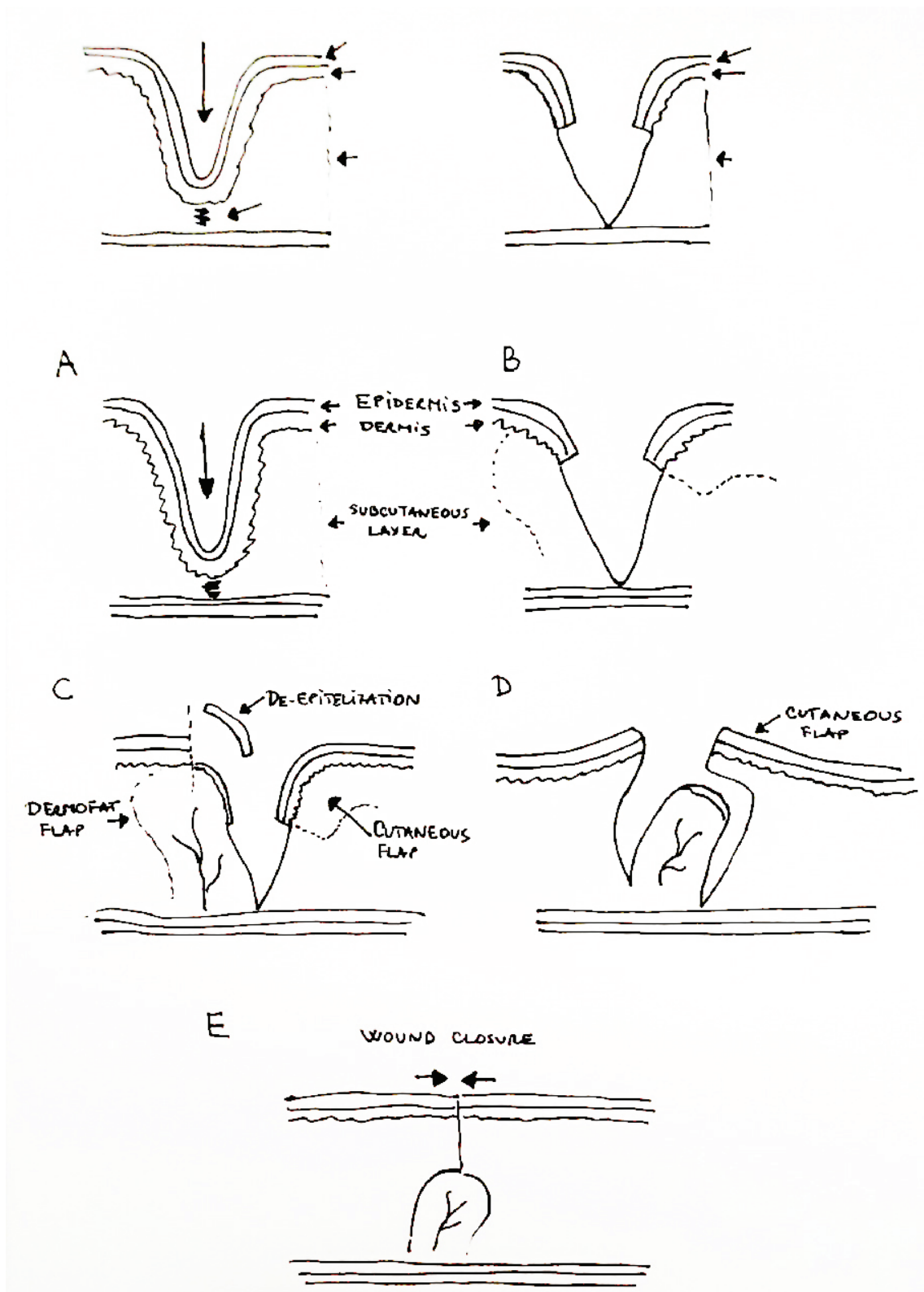
We performed surgery under general anesthesia with a tourniquet application. The scheme of the Mutaf technique is shown in **Figure 1**. Constriction rings were identified intraoperatively and carefully excised (all fibrotic tissue had to be excised) (**Figure 1a,b**). Rectangular flaps were designed on both sides of the groove. The height of these flaps was extended up to the border of the normal limb contour, and the width of each flap was about twice as big as its height. Flaps were de-epithelialized in an alternating pattern on each side while the opposite flaps stayed intact (**Figure 1c**). The de-epithelialized rectangular flaps were elevated as dermofat flaps and turned over from both sides toward the groove and sutured to the subcutaneous tissue of the opposing rectangular skin flaps (**Figure 1d**). According to this technique, the groove was filled with a soft tissue bracelet created by the turned-over dermofat flaps. The skin closure was obtained with rectangular cutaneous flaps opposing the dermofat flaps (**Figure 1e**).

## CASE REPORT 1

We present our first patient with CCR on the distal part of the right forearm with circumferential swelling of the right hand and arm distal to the constriction. According to Patterson, this was CCR II type (**Figure 2a**). Due to conspicuous lymphoedema and functional impairment of the hand, we decided on the one-stage Mutaf technique under general anesthesia. All neurovascular structures were identified and a constriction ring with all fibrotic tissue was excised. After excision, we designed rectangular turn-over dermofat flaps to fill the groove (**Figure 2b**). Ultimately, the skin closure was obtained with rectangular cutaneous flaps opposing the dermofat flaps. There were no problems with circulation or innervation in the postoperative period, and the functional and aesthetic outcome was satisfying (**Figure 2c**). We have continued follow-up for 6 years and here we present the result (**Figure 2d**).

## CASE REPORT 2

Our second patient had type III CCR with associated multiple foot and hand anomalies. Multiple constriction

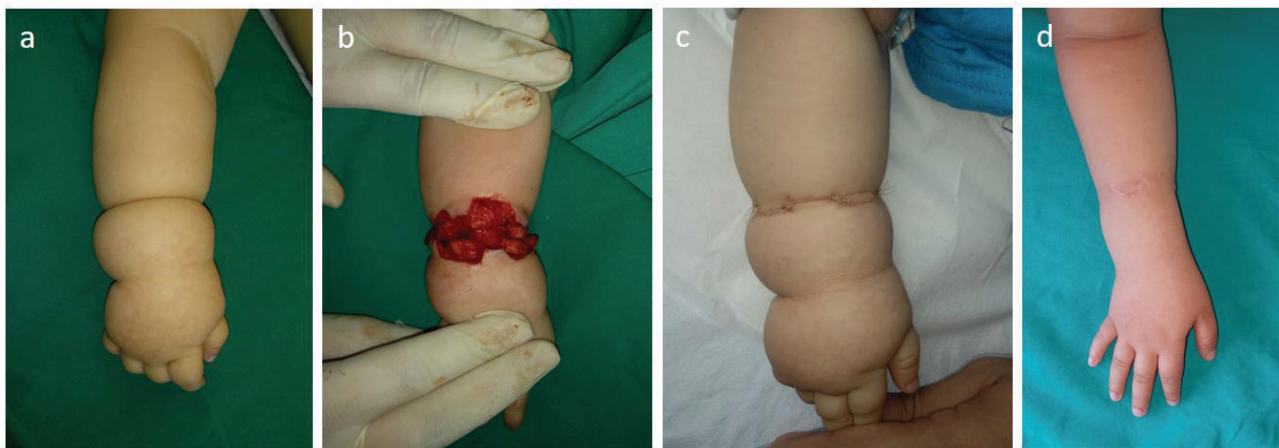


**Figure 1.** Schematic presentation of the one-stage Mutaf technique and its basic principles

- a. Congenital constriction ring;
- b. Excision of the ring;
- c. De-epithelization of the dermofat flap;
- d. Elevation of the dermofat as a cutaneous flap;
- e. Result

bands on the distal part of the right leg with clubfeet were present (**Figure 3a**). Due to vascular compromise and lymphoedema, we decided to perform the one-stage

Mutaf procedure. An extremely strong and thin constrictive ring was present and the preparation of the neurovascular structures was extremely delicate (**Figure 3b**).



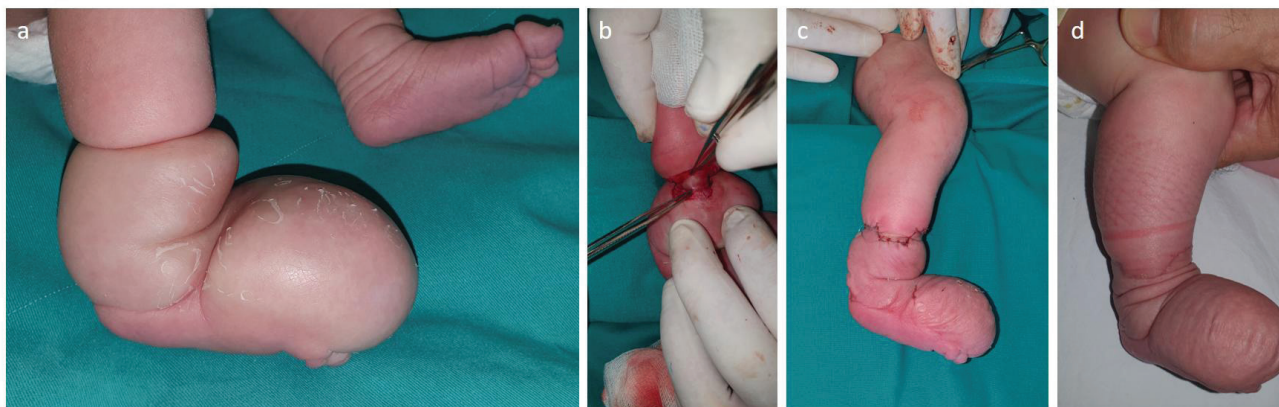
**Figure 2.** Case presentation of a patient with type II congenital constriction ring of the right arm  
 a. Congenital constriction ring of the right arm with distal lymphoedema  
 b. Intraoperative view with rectangular skin and dermofat flaps  
 c. Result after two weeks postoperatively  
 d. Result after six years of follow up.

Additionally, conspicuous lymphoedema was intraoperatively identified. We designed rectangular flaps according to Mutaf (Figure 3c). We got satisfying results immediately after the procedure, with no present neurovascular compromise. Postoperatively, excess skin was present on the dorsum of the right foot. Here we present results after six months (Figure 3d). The patient is still on compressive therapy, and further reconstructive procedures (skin reduction) of the right foot are planned.

**Discussion**

The time and the approach to the management of CCR depend on clinical presentation and range from fetal surgery and urgent postnatal surgery to multiple-stage surgeries performed later in life. (1,3,9,10,12,14,15,18,20) In cases where circulation or innervation is compromised, and where there is a risk of gangrene autoamputation surgery has to be performed as soon as possible.

(1,2,9,11,15) This was the case with our patient N<sup>o</sup>2. Some authors performed surgeries immediately after birth. (14,16) On the other hand, several studies showed very good results in patients who started with surgeries a few months after birth or even later. (3,7) In our report of the first case we had a lower risk of neurovascular obstruction, so we performed surgery at a later date, while the patient described in the second case had a high risk of neurovascular obstruction and surgery was performed a few days after birth. There are several options for the treatment of CCR such as multiple Z- or W- plastics, one-stage circular resection and primary circular suture without Z-plastics, simple linear hemi-circumferential excision without Z-plastic, rectangular-plastic technique (the one-stage Mutaf procedure), and multiple continuous opposing Y-to-V-plastics in a single or staged manner. (9-19) Regardless of the technique used, the key to success is a complete resection of all constricting rings and fibrous tissue, and linear incisions of the deep fascia. (1,



**Figure 3.** Case presentation of a patient with type III congenital constriction rings of the right leg  
 a. The large oedema of the distal part of right leg as a consequence of congenital constriction ring  
 b. Excision of fibrous band which cause the oedema  
 c. Immediate postoperative result with the Mutaf technique  
 d. Result after six months of follow up

-3,7,9,10,12-18) Straight circular sutures at the limbs are generally avoided because of possible scar contractures and circular constrictions, however, this technique is still used by some authors. (1-9,16,17,18) To reduce the risk of scar contracture some authors advocate multiple Z- or W-plastics and according to their experience transposition of two or more flaps allow additional relief of skin tension by redistribution of a relative excess of adjacent skin. (1-7,9-14,18) The main disadvantage of Z-plastic is the undesirable course of scars and the rest of contour deformity. (1) In our point of view using rectangular flaps according to the Mutaf technique can both reduce skin tension and obtain better aesthetic results than Z- or W-plastics.

In the CRS we make soft tissue deficiency up with similar tissue to obtain normal contour of limbs. The Mutaf technique advocates using well-vascularized turn-over dermofat flaps elevated in an alternating fashion from each side of the ring to fill the groove. (9) The incisions made this way are parallel to the relaxed skin tension lines. When the groove is filled rectangular skin flaps are sutured and suture lines are parallel to the relaxed skin tension lines.

Some authors advocate the two-stage “sine plastic” technique with a one-week interval between stages claiming that the known technique is not effective in the elimination of contour deformity. (16) In our point of view using the Mutaf procedure we can also achieve a similar aesthetic outcome in one stage resolving contour deformity as shown in Case 1. The benefits of one-stage procedures are a reduction in surgical and psychological trauma of patients and their parents, less time spent under general anesthesia and a lower amount of anesthetics the patient receives, and lower costs of hospitalization.

In the cases we present here, we achieve satisfactory results in correcting constriction rings in the arm and the

leg. Hence, we think that the Mutaf technique is very reliable when it comes to the correction of constriction rings in the upper and lower extremities. On the other hand, this technique does not seem to be feasible for correcting constriction rings located at the acral parts of the foot and hand because of the lack of dermofat tissue for harvesting adequate flaps. (9)

## CONCLUSION

We find the Mutaf technique to be an effective and safe one-stage technique for the treatment of congenital constriction rings.

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None

## Conflict of interest:

None to declare.

## Author Contributions:

All authors contributed to the study conception and design. Aleksandar Vlahović is in charge of the design, concept, main idea and the guarantor of integrity of the entire study. Milana Živković is in charge of the study concepts, design and manuscript preparation. Ninoslav Begović is in charge of the material preparation, analysis and editing. Ivan Dizdarević is in charge of the study design and manuscript preparation. Nataša Vlahović is in charge of the study concepts, data acquisition and manuscript preparation. All authors declare that the details of any figure can be published.

## REFERENCES

- Habenicht R, Hülsemann W, Lohmeyer JA, Mann M. Ten-year experience with one-step correction of constriction rings by complete circular resection and linear circumferential skin closure. *J Plast Reconstr Aesthet Surg*. 2013;66(8):1117–22. doi: 10.1016/j.bjps.2013.04.042 PMID: 23660282
- Homer LE, Mishra A, McArthur P. Amniotic constriction bands: a case series and proposed new classification system. *Hand Surg*. 2015;20(1):121–6. doi: 10.1142/S0218810415500173 PMID: 25609285
- Rossillon D, Rombouts JJ, Verellen-Dumoulin Ch, Vanwijck R, Vincent A, de Coninck A. Congenital ring-constriction syndrome of the limbs: A report of 19 cases. *Br J Plast Surg*. 1988; 41:270–7. doi:10.1016/0007-1226(88)90111-7 PMID: 3382854
- Adu EJK, Annan C. Congenital constriction ring syndrome of the limbs: a prospective study of 16 cases. *Afr J Paediatr Surg*. 2008;5(2):79–83. doi:10.4103/0189-6725.44182 PMID: 19858673.
- Ozkan K, Unay K, Goksan B, Akan K, Aydemir N, Ozkan NK. Congenital constriction ring syndrome with foot deformity: two case reports. *Cases J*. 2009; 2:6696. doi: 10.4076/1757-1626-2-6696 PMID: 19829844.
- Patterson TJ. Congenital ring constrictions. *Br J Plast Surg*. 1961; 14:1-31. doi: 10.1016/s0007-1226(61)80002-7 PMID: 13733379
- Walter JH Jr, Goss LR, Lazzara AT. Amniotic band syndrome. *J Foot Ankle Surg*. 1998;37(4):325–33. doi: 10.1016/s1067-2516(98)80070-7 PMID: 9710786
- Shetty P, Menezes LT, Tauro LF, Diddigi KA. Amniotic band syndrome. *Indian J Surg*. 2013;75(5):401–2. doi: 10.1007/s12262-012-0468-x PMID: 24426485
- Mutaf M, Sunay M. A new technique for correction of congenital constriction rings. *Ann Plast Surg*. 2006;57(6):646–52. doi: 10.1097/01.sap.0000235430.21875.55 PMID: 17122551
- Miura T. Congenital constriction band syndrome. *J Hand Surg*. 1984;9:82–88. doi: 10.1016/s0363-5023(84)80191-4 PMID: 6319481
- Kay SP, McCombe D, Kozin SH. Deformities of the hand and fingers. In: Green DP, editor. *Green's operative hand surgery*. Philadelphia: Elsevier Churchill Livingstone; 2005. p. 1381-444.
- Wang L, Fang Y. Clinical assessment of the results of one-stage circular incision techniques for limb ring constriction due to am-

- niotic band syndrome. *Medicine (Baltimore)*. 2021;100(30):e26764. doi:10.1097/MD.00000000000026764 PMID: 34397722
13. Upton J, Tan C. Correction of constricted rings. *J Hand Surg*. 1991;16(5):947-53. doi: 10.1016/s0363-5023(10)80166-2 PMID: 1940180.
  14. Prasetyono TOH, Sitorus ASN. A review on the safety of one-stage circumferential ring constriction release. *Int Surg*. 2015;100(2):341-9. doi:10.9738/INTSURG-D-13-00230.1 PMID: 25692440
  15. Chan AHW, Zeitlinger L, Little KJ. Multiple continuous Y-to-V-plasties for excision and reconstruction of constriction band syndrome: Case series and description of surgical technique. *Plast Reconstr Surg*. 2022;149(4):774e-8e. doi: 10.9738/INTSURG-D-13-00230.1 PMID: 25692440
  16. Hung NN. Congenital constriction ring in children: sine plasty combined with removal of fibrous groove and fasciotomy. *J Child Orthop*. 2012;6(3):189-97. doi:10.1007/s11832-012-0420-4 PMID:23814619.
  17. Tan P-L, Chiang Y-C. Triangular flaps: a modified technique for the correction of congenital constriction ring syndrome. *Hand Surg*. 2011;16(3):387-93. doi: 10.1142/S021881041100576X PMID:22072482
  18. Choulakian MY, Williams HB. Surgical correction of congenital constriction band syndrome in children: Replacing Z-plasty with direct closure. *Can J Plast Surg*. 2008 Winter;16(4):221-3. doi: 10.1177/229255030801600409 PMID: 19949501
  19. Payen M, Luc F, Sales de Gauzy J, Accadbled F, Abid A. Libérations des sillons de constriction amniotique par excision hémi-circonférentielle et fermeture cutanée sans plastie en Z. *Ann Chir Plast Esthet*. 2022; 67(2):73-80. doi: 10.1016/j.anplas.2022.01.004 PMID: 35183393.
  20. Minella C, Costantino B, Ruano R, Koch A, Weingertner AS, Favre R, Sananes N. Fetoscopic release of amniotic band syndrome: an Update. *J Ultrasound Med*. 2021;40(5):1039-1048. doi: 10.1002/jum.1548 PMID: 32951245.

## JEDNOAKTNA PROCEDURA PO MUTAFU ZA UROĐENE KONSTRIKTIVNE PRSTENOVE - BEZBEDNA I EFIKASNA

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### Sažetak

**Uvod:** Sindrom amnionskih brida predstavlja retku kongenitalnu malformaciju koja najčešće zahvata ekstremitete, ređe glavu i trup. Nastaje zbog fibroznih brida koje se tokom intrauterinog razvoja obmotavaju oko ploda, pri čemu može doći do akrosindaktilije, kompresije na ekstremitetima, sa ili bez kompromitovanja vaskularizacije, ili u najtežim slučajevima do amputacije različitih delova ekstremiteta. Postoji nekoliko hirurških tehnika za rešavanje ove anomalije. U ovom radu, prikazali smo naše rezultate koristeći tehniku po Mutaфу u jednom aktu.

**Prikaz pacijenata:** Prikazali smo dva deteta sa sindromom amnionskih brida. U prvom slučaju, prikazano je dete sa konstriktivnim bridama na desnoj ruci sa izraženim limfedemom i deformitetom distalno od mesta kompresije. U drugom slučaju, prikazano je dete sa multiplim konstriktivnim bridama na distalnim delovima desne potkolenice sa deformitetima stopala i izraženim

limfedemom uz kompromitovanje vaskularizacije. U oba slučaja primenjena je Mutafova tehnika u jednom aktu u opštoj anesteziji. U cilju obezbeđivanja normalne konture ekstremiteta, dizajnirani su pravougaoni "turn-over" režnjevi potkožnog masnog tkiva. Za rekonstrukciju defekta kože, korišćeni su pravougaoni kožni režnjevi. U oba slučaja postoperativni period je protekao uredno. Postignut je zadovoljavajući estetski rezultat. Sa obzirom na to da se radi o tehnici koja se izvodi u jednom aktu, najveća prednost ogleda se u manjem stresu kod roditelja i deteta, kraćem trajanju anestezije i manjim troškovima hospitalizacije u odnosu na multiple procedure.

**Zaključak:** Mutafova tehnika predstavlja efektivnu i bezbednu tehniku za rešavanje sindroma amnionskih traka u jednom aktu, kojom se postiže normalna kontura ekstremiteta bez limfedema, uz zadovoljavajući izgled ožiljka.

**Ključne reči:** Sindrom amnionskih traka, hirurgija, tehnika

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