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One stage release of circumferential constriction band in an upper limb with multiple Z-plasty techniques: a case report



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ABSTRACT

Background: Congenital constriction band is an uncommon congenital abnormality with multiple disfiguring and disabling manifestations. The widely known management for the correction of congenital constriction band especially in circumferential constriction band is done in a multistage release of constriction band to eluding vascular and lymphatic embarrassment. The purpose of this case report was to present the outcome of one-stage release in terms of safety and aesthetic appearance.

Case Description: Women 28-year-old came to the outpatient plastic surgery clinic presented with circumferential constriction band with no functional disability in her right arm since birth. Excision based on the design of multiple Z-Plasty is started 2 cm proximal and distal from the constricted band. The multiple Z-Plasty flaps are transposed around each other, changing the direction of the constriction band. Postoperatively, the extremity circulation was

normal, and no edema was observed. The range of motion is within normal limits and aesthetically acceptable for the patient. Patterson classified constriction ring syndrome into four types, and type II is the most common. In type II Paterson, there are normal full-layer skin structures, and no important nerve or blood vessel was affected so an incision completely encircling an extremity would not interfere with distal circulation and provided strong evidence that one-stage resection of constriction band could be performed. Using multiple Z-Plasty for constriction band release leads to improvement of the functional and cosmetic appearance of the defect.

Conclusion: A one-stage release of circumferential constriction band in the upper limb using multiple Z-Plasty is considered as a safe treatment with an aesthetically acceptable result for the patient

Keywords: Constriction Band, One-stage release, Multiple Z-Plasty

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INTRODUCTION

Constriction band syndrome also known as constriction ring syndrome, amniotic disruption sequence or amniotic band syndrome is characterized by a fibrous band that tightly encircles the limb, either partially or entirely.¹ The incidence ranges from 1 in 1200 to 15,000 live births, which the ratio between male and female affected is approximately 1:1.^{2,3} No evidence of familial hereditary tendency has been found.¹ There are 3 theories that widely known among hand and plastic surgeons as the pathogenesis of constriction band syndrome; they are intrinsic theory, extrinsic theory, and intrauterine trauma theory.^{4,5} Patterson introduced a classification of constriction band syndrome into four types, the first type is simple constrictions only, the second type is when constrictions with distal deformity, the third type is constrictions with a fusion of distal parts; and the fourth type, constriction cause intrauterine amputations.⁵ This classification system is based on the severity of the syndrome and is useful because

the different types require different treatments.

The concept advocated by Stevenson was to release the constriction ring in 2 or 3 stages usually with multistage Z or W-Plasty to prevent circulatory and lymphatic disturbance.^{6,7} Several modifications on the management of congenital constriction band have been attempted, one of them is the one-stage circumferential constriction band release with Z-Plasty. A one-stage release of congenital constriction band results in one-stage correction avoids repeat surgery, requires easy postoperative care, reduces the financial burden, and saves patient's time.⁷ Case of one stage circumferential constriction band release using multiple Z-Plasty in the adult is presented.

CASE DESCRIPTION

A Female 28-year-old came to the outpatient plastic surgery clinic presented with a circumferential constriction in her right arm since birth. There is no functional disability in her arm. No family history of hand and foot deformity. Physical examination



Figure 1. Circumferential constriction band in the right arm with normal humerus, radioulnar bone, and elbow joint



Figure 2. Design of multiple Z- plasty in which excision started 2 cm proximal and distal from the constricted band

shows there is no tenderness in the constricted part and range of motion of elbow flexion and extension is within normal limit. There was no discoloration of the skin and no enlargement of superficial veins. There is no amputated part of her fingers. General physical examination was within normal

limits. From radiological examination shows no abnormality of elbow joint, humerus and radioulnar bone (Figure 1).

The design of multiple Z-Plasty was used for the surgery as shown in Figure 2. In this procedure, the contracture band is divided into several segments, each with a Z-Plasty designed in series. After preparation of the part, marking ink is applied to the band circumferentially to assess the apposition after the release. This marking ink gives the surgeon an idea about the amount of skin to be excised from both sides of the band. Excision based on design is started 2 cm proximal and distal from the constricted band (Figure 2).

Full-thickness skin flaps are undermined at the level of subcutaneous fat, creating numerous flaps of equal size and shape. An adequate undermining of surrounding subcutaneous tissue is performed to achieve proper mobilization of the flaps. The multiple Z-Plasty flaps are then transposed around each other, changing the direction of the constriction band. The flaps are held in place with a few anchoring stitches. The skin is closed using interrupted sutures. Topical antibiotics and a pressure dressing are applied over the wound to minimize the chance of hematoma formation caused by the undermining. Circulation to the leg and foot are assessed by capillary refill and color of the skin. Postoperatively, the part is massaged distal to proximal to reduce edema. The wound is re-examined in one to two days, and sutures are removed in 14 days. The outcome was satisfactory as shown below (Figure 3). The extremity circulation was normal, and no edema was observed postoperatively. There is no functionality problem in her arm; the range of motion is within normal limit.

DISCUSSION

Few studies have investigated the treatment strategies for congenital constriction band syndrome. The effects on the blood flow are one of the major concerns that restrict the surgical treatment of congenital constriction ring syndrome. The most commonly used surgical method for this disease is multistage Z-Plasty, which releases the constriction band in multiple stages to prevent circulatory disturbance. However, this treatment involves multiple operations that result in surgical and psychological trauma, increase amount of anesthetic the patient takes, and also the time of hospitalization, and the cost. This multistage treatment also associated with a relatively high recurrence rate and poor postoperative appearance.¹

Based on researches,⁸⁻¹⁰ one-stage release of constriction band does not place the underlying



Figure 3 Postoperative result of one stage release of the constriction band with multiple z plasty. No vascular compromise and no edema observed postoperatively.

neurovascular structures at greater risk than multiple staged Z-Plasty. There was no skin necrosis or wound healing problem. Further studies on circulation to the skin flaps found that blood supply to the skin is primarily from the musculocutaneous arteries that directly penetrate the subcutaneous and cutaneous tissue from underlying muscles.^{7,11} This is very important to support one stage contracture release in constriction band.

Patterson classified constriction ring syndrome into four types, and type II is the most common, referring to a constriction ring with distal deformity, including atrophy and lymphedema.¹ One stage release of constriction band in 21 consecutive patient with constriction band syndrome type

II shows great result with less morbidity.¹ This study conducted with Jiang et al. showed that the constriction rings in patients with Patterson type II congenital constriction ring syndrome consisted of normal full-layer skin structures with complete basement membrane and no detachment between the epidermis and dermis.¹ Subcutaneous collagen with high-density aggregates were found.¹ The subcutaneous tissue space was normal, and no important nerve or blood vessel was affected.¹ These findings show that an incision completely encircling an extremity would not interfere with distal circulation, and provided strong evidence that one-stage resection of constriction band could be performed.^{1,12}

One stage release of constriction band is not necessarily applicable with Patterson type III or higher congenital constriction band syndrome, those with severe edema of the distal extremity, and those with blood flow disorders. More studies still need to be conducted related to the management of these patients.

The functional and aesthetic appearance produced by constriction ring syndrome is proportional to the degree of severity. Treatment must provide not only good results functionally but also an excellent aesthetic appearance because, in most cases, aesthetic appearance is usually the main reason for consultation. Z-Plasty technique is one of the most versatile and widely used maneuvers in the plastic surgery that surprisingly simple, yet incredibly effective.^{13,14} Z-Plasty release contractures by increasing the length along the axis of the scar and breaking up the straight-line traction of the scar that leads to improvement of the functional and cosmetic appearance of scars.¹³ Multiple Z-Plasty in series is one the modification of Z-Plasty which the contracture band is divided into several segments, each with a Z-Plasty designed in series. It seems obvious that using multiple small Z-Plasty in series is better than using one large Z-Plasty because the need for shortening width to give increased length is less when the Z-Plasty is smaller.¹⁵ In this case report, scar after multiple Z-Plasty procedures is still aesthetically acceptable for the patient.

CONCLUSION

This case report confirms that one-stage release of circumferential constriction band in the upper limb using multiple Z-Plasty is considered as a safe treatment with an aesthetically acceptable result for the patient. This technique avoids repeat surgery, requires easy postoperative care, reduces the financial burden, and saves patient's time.

CONFLICT OF INTEREST

The authors declare that there is no competing interest regarding the manuscript

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AUTHOR CONTRIBUTION

IGPHS and ARHH are responsible for the surgical technique used and clinical outcome assessment in this study as well as a conceptual framework of manuscript preparation. GWS is responsible for English improvement and assisted the prior two authors in the operating room.

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