Modified Z-Plasty Repair of Webbed Neck Deformity Seen in Turner and Klippel-Feil Syndrome

Objective: Webbed neck deformity exists in many syndromes including Turner's or Klippel-Feil syndrome. Multiple problems are encountered with existing techniques to correct a webbed neck deformity. In Turner's syndrome, a subcutaneous band of thickened fascia and a low neck hairline present a challenge to the surgeon when designing a repair. The authors propose the following new technique that addresses both issues.

Material and Methods: Five patients with webbed neck underwent this new procedure. A Z-plasty is performed with the midline arm down the length of the web. The subcutaneous fibrous band is excised, the shortened trapezius is released, and the hair-bearing flap is excised. The anterior flap is rotated and advanced to join its mate flap from the contralateral neck at the posterior midline. A resultant dog-ear near the acromion is corrected with an additional Z-plasty.

Results: In all five patients, the functional and aesthetic results were very satisfactory to both patient and surgeon. An 11-year follow-up is presented with excellent correction of the webbing. Both limited range of motion and the cosmetic deformity are addressed by this technique.

Conclusion: The results obtained by using the simplified modified Z-plasty technique for repair of webbed neck deformity are very satisfactory. We propose the use of this technique for correction of webbed neck deformities whenever the posterior surface of the neck web contains a significant amount of hair.

KEY WORDS: Klippel-Feil syndrome, Turner's syndrome, webbed neck, Z-plasty

Webbed neck deformity is an anomaly associated with a number of syndromes. It is a defect with numerous pathways of genetic transmission and has a variety of methods of surgical correction. In 1912 Klippel and Feil described a webbed neck condition associated with cleft palate, low posterior hairline, and restricted cervical motion. This immobility was painless and secondary to variable fusion of the cervical vertebrae (McCarthy, 1990). Turner (1938) reported a series of female patients who exhibited sexual infantilism, webbed neck, short stature, and low posterior hairline. Patients with this condition (Turner's syndrome) genotypically lack the second X chromosome in the 23rd pair (Castiglia, 1997).

The variety of subcutaneous structural anomalies associated with these syndromes can affect the repair of the webbed neck. The depth of range of motion is of less importance to the patient, however, than is the cosmetic deformity. In Turner's syndrome, there is most often a subcutaneous band of thickened fascia running from the mastoid to the acromion process, sometimes with muscle involvement (Thomson et al., 1990). An adequate repair can be facilitated by fascial band excision through a variety of approaches. The most difficult obstacle in these patients is to overcome the poverty of skin and subcutaneous neck tissue and shortened muscle and fascia.

Many techniques have been described to correct a webbed neck deformity. With a midline posterior advancement flap (Thomson et al., 1990), not only is the hairline elevated, but also much of the scar is hidden within the hairline. This is accomplished with a suture line closure that runs caudal to cranial with excision of the dog-ears created at the superior-Y shaped wound margin. The web is obliterated by removal of the excess skin posteriorly with closure of the defect so as to flatten the web against the side of the neck. Unfortunately, this scar is under considerable tension and is likely to expand over time. There is a tendency for the web to return (Miller et al., 1990).
Shearin and Defranzo (1980) described a midline posterior butterfly incision in which the loose skin and subcutaneous tissue was pulled to a midpoint on the back of the neck. Although this approach suppressed the web immediately postoperatively, the fibrous band was never excised and the web reappeared 3 months later.

Menick et al. (1984) reported a lateral cervical advancement flap that elevated the platysma with the anterior neck skin and used posterior, superior retraction to obliterate the fibrous web. This technique had the advantages of elevating the low hairline and allowing direct visualization of the structural anatomy of the web, but the fibrous band of fascia was never excised (Thomson et al., 1990).

Expanders could ideally create more hairless skin and subcutaneous tissue in the vertical plane; however, they have failed to achieve this goal. Instead, the thick, hair-bearing skin on the posterior portion of the web migrated onto the expanders as they were inflated. It was found that the same degree of skin advancement could be achieved more simply by extensive undermining into the anterior cervical triangle. Another repair described a lateral approach that utilized a full-thickness skin graft. The cosmetic result was much less than was desired (Chandler, 1937).
Z-plasty to treat webbed neck deformity was first described in 1937 by Chandler. Many have subsequently tried the Z-plasty and have found the results unsatisfying. The Z-plasty has since been mentioned in the literature only for its drawbacks, in particular, the amount of hair-bearing skin left on the lateral neck. It is still the only technique described in the literature that fully augments the paucity of skin and subcutaneous tissue in the lateral web.

The Z-plasty approach, which is well described by MacGregor and McGregor (1995), has been modified and used by us for cosmetic and functional improvement of webbed neck caused by Turner’s and Klippel-Feil syndromes. The results were quite satisfactory.

**MATERIALS AND METHODS**

In an effort to improve on the usual Z-plasty, a modified Z-plasty was developed. Five patients with webbed neck underwent this new procedure. The patient was placed in prone position for bilateral repair. The fibrous band running from the mastoid to the acromion process was placed under tension and a Z incision made with the inferior arm toward the anterior neck, the middle arm down the length of the web, and the superior arm toward the posterior neck (Fig. 1). This was deep-
FIGURE 8  Postoperative lateral view of modified Z-plasty repair.

FIGURE 9  Postoperative posterior view of modified Z-plasty repair.

FIGURE 10  Eleven-year postoperative view of modified Z-plasty repair.
FIGURE 11 Eleven-year follow-up of modified Z-plasty repair (close-up view).

ened, and a large triangular flap based on the area of the scalp just behind the lower half of the ear was reflected posteriorly. The large anterior neck flap was then reflected forward. As expected, muscle and fascia were both contributing to the neck contracture. The superficial band of shortened fascia was excised down to the level of muscle, the anterior flap was undermined well into the anterior neck region, and the posterior flap was undermined to the midline of the posterior neck (Fig. 2). Attention was then turned to the trapezius border contributing to the web. With a wet sponge laid over the top of the muscle to compress and contour it, relaxing incisions were made in the superior and inferior contracted margins of the trapezius. With the help of these transverse relaxing incisions, the muscle lay easily into the neck contour (Fig. 3). At this point, the head could be deflected to the opposite side an additional 20 degrees. The desired hairline from mastoid to posterior midline could then be drawn. An incision was made along this hairline so as to excise and discard virtually all of the heavy hair-bearing skin comprising the triangular, posterior flap of the Z-plasty (Fig. 4). The anterior flap was then advanced and brought backward to join, at the midline of the neck, its mate flap. This mate flap had been elevated from a similar large Z-plasty of the web on the opposite side of the neck. Two smaller zigzag flaps were then fitted into the neck laterally near the acromion to remove as much of the remaining hair-bearing skin as possible (Fig. 5). Very little excess skin had to be trimmed away in the final dog-ear on the lateral neck.

RESULTS

All five patients experienced improved cosmetic appearance and noticeable increase in range of motion. Preoperatively, the mother of the 3-year-old patient expressed concern for her daughter’s obvious web (Figs. 6 and 7). Shown postoperatively, the issues of cosmesis and range of motion had both been addressed (Figs. 8 and 9). Follow-up 11 years later demonstrated functional and aesthetic results that were satisfactory to both patient and surgeon (Figs. 10 and 11).

CONCLUSION

There are many corrective procedures for webbed neck that give improved cosmetic and functional outcomes. The plastic surgeon constantly searches for the method that gives the maximum result and the minimum morbidity. The standard Z-plasty technique has been abandoned by so many for webbed neck repair because of the subsequent anterolateral scars that are visible in the anterior neck and for the remaining, unsightly hair-bearing tissue that is displaced forward. The authors believe the benefits of the modified Z-plasty repair to be greater than any other of the procedures described. The fibrous band that is the scaffold for the web is easily excised under direct visualization, and the shortened muscle fibers of the trapezius muscle may then be gently stretched or even myomtomized to normalize the curvature of the lateral neck. The anterior flap of each of the two large Z-flaps may have its postoperative scar exactly placed along the edge of the hairline in each postauricular region where it will be covered by hair and lengthens the congenital skin shortage. The Z-plasty variation described here avoids displacement of hair-bearing tissue onto a visible area in the lateral neck. Future studies directed at laser resurfacing of visible scars combined with postoperative laser removal of any residual undesirable fine hair is sometimes a useful adjuvant to this modified Z-plasty approach to web neck deformity. The excision of the hair-bearing Z-plasty flap obviates the need for laser or electrolysis hair removal treatments. We propose the use of this technique for the correction of
webbed neck deformities whenever the posterior surface of the neck web contains a significant amount of hair.

REFERENCES


