

## CASE REPORTS

### SURGICAL CORRECTION OF A WEBBED-NECK DEFORMITY IN TURNER'S SYNDROME

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Turner's syndrome occurs in approximately 1 out of every 2,000 to 2,500 live-born girls. This genetically determined pathology is characterised by multiple congenital anomalies. A typical form of this syndrome is associated with a lack of one of the sex chromosomes (karyotype 45, X). From the point of view of plastic surgery, one of the most important elements of the therapy is correction of the webbed neck deformity.

The aim of the study was to present the possibilities of surgical treatment of a webbed neck of patients with Turner's syndrome and the evaluation of treatment results.

In the years 2000–2012, six children with Turner's syndrome were treated because of the webbed neck deformity. The age of patients ranged from 9 to 17 years. In the case of all patients, the aim was to distribute the neck skin folds by using Z-plasty in conjunction with a shift to the back of glabrous skin flaps mobilised from the anterolateral surface of the neck.

In the case of four operated patients, the folds were completely removed and a correct symmetrical outline of the neck was obtained. One patient was found to have unilateral moderate webbed neck recurrence after about 2 years of treatment. In one case, the correction was insufficient. The performed surgical procedures enabled correction of low hairline only in the lateral parts of the neck. The lower line of the scalp in the central part of the neck has remained unchanged.

The lateral approach with a shift of glabrous skin flap to the back, which we performed, allows for effective reduction of the webbed neck, excision of bands of the connective tissue and correction of the low hairline on the side of the neck. Z-plasty enables an adequate extension of scars and improves the contour of the neck.

**Key words:** web neck deformity, Turner's syndrome, surgical correction

Turner's syndrome occurs in approximately 1 out of every 2,000 to 2,500 live-born girls. This genetically determined pathology is characterised by a number of congenital disorders (short height, broad chest with widely placed nipples, gonadal dysgenesis, primary amenorrhea, aortic coarctation, bicuspid aortic valve, congenital kidney abnormality, facial cranium defects) (1, 2, 3). A typical form of this syndrome is associated with a lack of one of the sex chromosomes (karyotype 45, X), but mosaics (e.g. 45,X/46,XX or 45,X/46,XY) are also fairly often, as well as structural abnormalities of chromosome X (4).

Turner's syndrome is increasingly being diagnosed prenatally or in infancy or early childhood, which is crucial for the quick implementation of multidisciplinary (both pharmacological and surgical) treatment of the disease (5). Turner's syndrome treatment methods are constantly developing (6). A milestone in the treatment of this syndrome was the introduction of an estrogen replacement therapy in the first half of the last century by an American endocrinologist Henry Turner. The latest scientific reports seem to confirm the positive effects of the use of a growth hormone in the early stages of treatment (7). From the point

of view of plastic surgery, one of the most important elements of the therapy is correction of the webbed neck deformity. Such abnormality is also noticed in other congenital malformation syndromes (e.g. in Noonan syndrome or Escobar syndrome). In Turner's syndrome, the webbed neck is accompanied by a low hairline, which is an additional difficulty while planning a surgery (8).

A webbed neck, occurring in approximately 15 per cent of patients with Turner's syndrome, is probably a remnant of the backflow of subcutaneous lymphatic obstruction around the neck formed during fetal development. Extensive subcutaneous cysts that form on both sides of the neck, cause stretching of the neck skin in the initial period and then, as a result of their sclerosing, shrinking and scarring, even folds are formed on the lateral surfaces of the neck, extending from the mastoid processes of the temporal bone to the coracoid process of the shoulder blade (9).

A webbed neck in patients with Turner's syndrome rarely causes functional limitation of movement, but it is a serious aesthetic problem as the deformity is difficult to hide. This pathology leads to the isolation of the girls from the peer group, which significantly worsens their emotional development. A correction of this distortion contributes to the child's better acceptance by peers, which means that the child may grow up with lesser fear of rejection (10).

The aim of the study was to present the possibilities of surgical treatment of a webbed neck in patients with Turner's syndrome and the evaluation of treatment results.

## MATERIAL

In the years 2000-2012, six children with Turner's syndrome were treated because of the webbed neck deformity. The age of patients ranged from 9 to 17 years (average 12 years). Before the operation, in the course of a clinical trial, the local disorders in patients were assessed, with particular regard to the extent of skin folds and skin excess within them, the course of hairline and occurring functional disorders.

The procedure was performed under general anaesthesia with endotracheal intubation in position on the side. In two cases a one-stage

procedure was performed on both sides of the neck, while the other four cases involved a two-stage procedure with 4-8 months interval between the operations.

In all patients, the aim was to distribute the neck skin folds by using Z-plasty in conjunction with a shift to the back of glabrous skin flaps mobilised from the anterolateral surface of the neck.

The incision was planned along the fold, on the border of the scalp. During the operation front hairless patches of the skin were widely dissected and they were slid back as far as possible, which allowed the excision of the scalp and covering the loss with hairless skin. In the next stage, in order to improve the contour of the neck, the scarring bands of connective tissue extending inside the folds were excised. In two cases, a surface myotomia of the lateral edge of the trapezius muscle was performed. Z-plasty was performed in order to extend the scar and improve its distribution. The patches with scalp produced here were maximally reduced in order to avoid scalp moving forward. The wound was sutured in layers, leaving a suction drain (fig. 1).

Follow-up examination were performed in all the operated children between 2 and 10 years after the operation. During the follow-up



Fig. 1. The condition after redistributing a skin fold

examination the following aspects were evaluated: the degree of correction of folds and the course of the hairline around the neck, the appearance of scars, the level of restriction of neck mobility and sensory disturbances. The patients were also asked for their own assessment of operation results.

## RESULTS

In four operated patients, folds were completely removed and a correct symmetrical outline of the neck was obtained (fig. 2AB, 3AB). One patient was found to have a unilateral moderate webbed neck recurrence after about 2 years of treatment. In one case, the correction was insufficient. However, the patient was satisfied with the result and did not consent to the proposed reoperation. The performed surgeries enabled correction of a low hairline only in the lateral parts of the neck. The lower line of the scalp in the central part of the neck has remained unchanged. However, the deformation in this area is not a problem for patients as it is easy to conceal with hair. In two girls, the follow-up examination revealed hypertrophied scars, which were treated conservatively, eventually achieving a good result. There were no limitations of the mobility of the neck or sensory disturbances in the operated area. Four patients found the result of the operation very good, while the other two found it good. The cause of the worse evaluation by one of them was insufficient

reduction of the folds, whilst the other one assessed that the correction of hairline around the neck was inadequate.

## DISCUSSION

A corrective surgery of a webbed neck was first described by Chandler, who used multiple Z-plasty in order to extend and distribute a skin fold on the anterolateral part of the neck (11). The main disadvantage of this method is visible scars and moving the scalp to the anterolateral surface of the neck. To minimise this, Hikade et al. proposed the implementation of a single wide Z-plasty. They planned the incision along the fold so that the middle ray of the Z-plasty went along the fold, on the border of the scalp. The result is the production of two flaps: one bottom pedicle flap containing scalp and the other upper pedicle flap containing hairless skin. The next stage of this technique is to shift the glabrous skin flaps to the back so that they “meet” each other in the midline on the back of the neck, and to arrange, fit and remove excessive hairy parts of the rear patches. Thus, hairy skin is not moved to the anterolateral surface of the neck (12).

Menick proposed correction of the webbed neck with lateral access. This technique involves mobilisation of a large flap on the anterolateral surface of the neck and moving it to the back. The incision is performed on the border between the front – hairless, and the



Fig. 2A. A 9-year-old patient with a webbed neck



Fig. 2B. The condition 10 years after the surgery



Fig. 3A. A 6-year-old patient with a webbed neck



Fig. 3B. The condition after the surgery at the age of 13

rear – hair-covered part of the fold, along its entire length. By mobilising the hairless skin together with the wide neck muscle, myocutaneous flaps are obtained, and after moving them upwards and backwards, they are used to cover losses created after the excision of excess scalp from the back of the neck fins (13).

A drawback of the method proposed by Menick is the fact that after adjusting one side, the opposing side is being stitched under some tension, which may cause a tendency for the skin to gradually stretch and, consequently, the webbed neck to recur. To avoid this problem in some patients we performed a two-stage procedure in 4-6 month intervals.

The method we used combines elements of operations proposed by Menick and Chandler. We found that the mere dissection of a skin flap on the front of the neck and its back shift does not allow a good correction of the skin fold. In order to achieve this, it is necessary to

extend the skin fold through the use of Z-plasty.

In order to avoid scars on the side of the neck, Foucar and then Shearin and DeFranzo described a webbed neck correction method involving a posterior approach. On the posterior surface of the neck, they excised a portion of the skin in the shape of a butterfly, and then dissected the skin around the wound edges and mobilised it towards the centre of the cavity, closing the wound in the shape of a double inverted letter "Y". A vertically extending part of the scar was, however, stretched to approximately 2.5 cm, due to which after about three months the deformation recurred (14).

In order to achieve lasting and positive results in the surgical treatment of the webbed neck, additionally to the reduction of excess skin and bottom hairline correction, it is very important to excise scarring bands of connective tissue located in the skin folds. Precise excision of the tensioning bands is possible only with the lateral approach. As described by Sherinie and DeFranzo, in the case of the posterior approach the scar tissue is not excised, which can cause the recurrence of the deformation observed by these authors. As we found extensive tensioning bands of connective tissue within the folds in all surgical patients, we did not apply the method proposed by these authors.

Among our patients undergoing surgical procedures, hypertrophic scars were found in two cases (33%), which is consistent with the results of other authors who observed it in about half of patients undergoing surgical procedures.

In conclusion, we can say that the choice of a surgical technique depends on the size of the skin folds, location of the hairline and the presence of bands of connective tissue. The lateral approach with a shift of glabrous skin flap to the back, which we performed, allows for effective reduction of the webbed neck, excision of bands of connective tissue and correction of the low hairline on the side of the neck. Z-plasty enables an adequate extension of scars and improves the contour of the neck.

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