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Staged Surgical Management of Turner Syndrome in a Pediatric Patient: a 5-year Longitudinal Follow-up

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ABSTRACT

Background: *Turner syndrome (TS) is a rare genetic condition characterized by complete or partial X-chromosome monosomy. It is associated with distinct phenotypic features, most notably webbed neck (pterygium colli) and ocular abnormalities such as medial epicanthus. Surgical correction of these conditions is technically challenging and associated with high recurrence rates, particularly in the pediatric population.*

Case Report: *We report the case of a 9-year-old girl with TS and multiple comorbidities, including congenital heart disease and a history of sequential surgical interventions since age 4. The patient underwent cervical and ocular reconstruction through serial Z-plasties and rotation flaps.*

Discussion: *The 5-year longitudinal follow-up demonstrates that a staged surgical approach, integrated with multidisciplinary care (endocrinology and cardiology), is essential to manage growth-related changes and systemic risks.*

Conclusion: *Long-term monitoring highlights the importance of individualized surgical planning and the adaptation of techniques throughout the patient's development to optimize functional and aesthetic outcomes, while mitigating the psychosocial impact of the syndrome.*

Keywords

Turner Syndrome, Pterygium Colli, Epicanthus, Surgical Flaps, Plastic Surgery, Pediatric, Z-plasty.

Introduction

Turner Syndrome (TS) is a rare genetic disorder caused by the complete or partial absence of one X chromosome, affecting approximately 1 in 2,500 live female births. Its clinical presentation is heterogeneous [1], characterized by short stature, gonadal dysgenesis, and a distinct phenotype that includes *pterygium colli* (webbed neck) and ocular abnormalities like medial epicanthus [2,3].

The surgical management of these deformities is complex, as it involves areas of high aesthetic and functional tension. In the pediatric population, the challenge is amplified by the risk of recurrence during growth and the presence of systemic comorbidities, such as congenital heart disease. This report presents the longitudinal follow-up of a patient who underwent sequential surgical corrections, emphasizing the importance of a multidisciplinary approach [1,4,5].

Case Report

A 9-year-old female patient, previously diagnosed with Turner Syndrome (45,X karyotype), was referred to the Plastic Surgery service. Her medical history was significant for congenital heart disease (interatrial communication - IAC), autism spectrum disorder (ASD), ADHD, and an allergy to fentanyl.

Physical examination revealed characteristic syndromic facies

(Figure 1), bilateral medial epicanthus (Figure 2), bilateral pterygium colli (Figure 3), lymphedema of the feet (Figure 4), and typical palatal abnormalities (Figure 5).

The patient's surgical history began at age 4 with an otoplasty for prominent ears, which resulted in a partial recurrence over the following years. At age 9, physical examination revealed a classic TS phenotype: low posterior hairline, bilateral *pterygium colli*, and bilateral medial epicanthus.

Figure 1: Nine Years Preoperative frontal view showing the characteristic facies of TS.

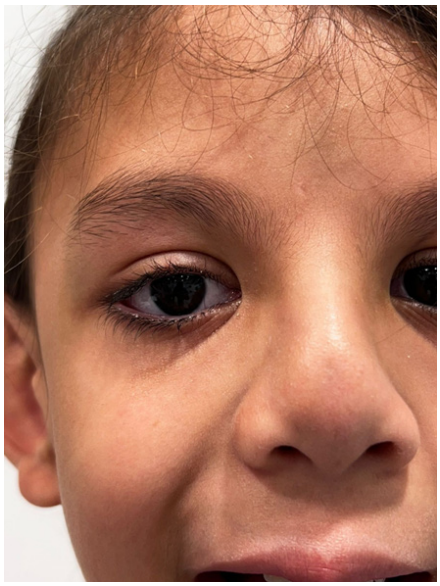

Figure 2: Preoperative close-up view showing medial epicanthus characteristic of TS.

Figure 3: Nine Years Preoperative view of the neck showing bilateral pterygium colli in a patient with TS

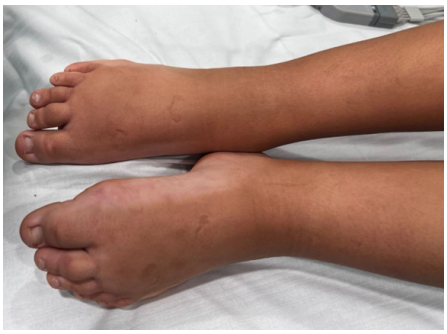

Figure 4: Edema in the feet, characteristic of Turner syndrome.

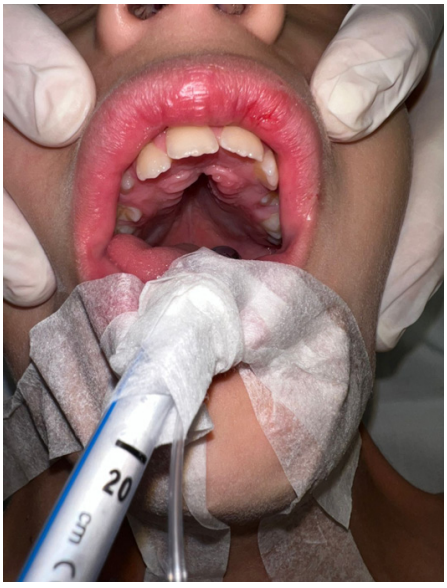

Figure 5: Intraoperative view of the oral cavity, showing the palate and dental arch in a patient with Turner syndrome.

Due to her clinical complexity, a staged surgical plan was established. The first stage involved the correction of the *pterygium colli* using bilateral serial Z-plasties to increase the vertical length of the cervical skin and redistribute tension (Figures 6,7). In the second stage, the medial epicanthus was corrected using bilateral rotation flaps to improve the palpebral contour and facial symmetry (Figure 8).

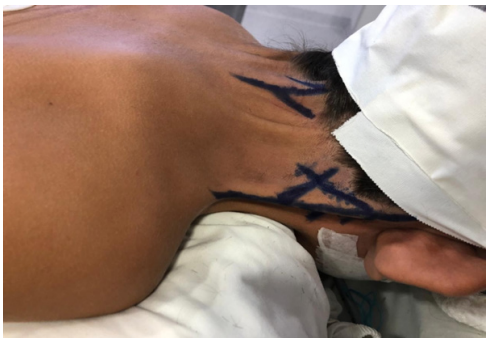

Figure 6: Preoperative marking of cervical Z-plasties for correction of pterygium colli in a patient with Turner syndrome.

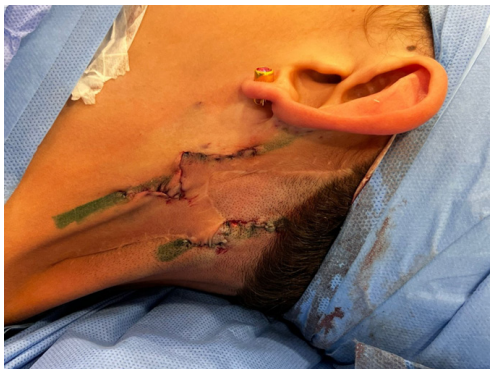

Figure 7: Immediate postoperative result of cervical Z-plasties for correction of pterygium colli in a patient with Turner syndrome.

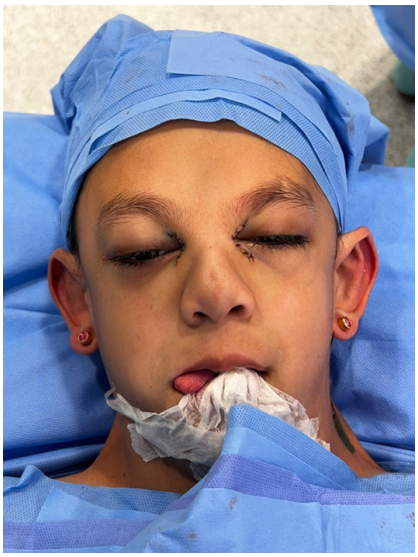

Figure 8: Immediate postoperative result of bilateral medial epicanthus correction with rotation flaps in a patient with Turner syndrome.

The procedures were performed under general anesthesia, with a specific protocol avoiding fentanyl due to her allergy. There were no intraoperative complications. The 5-year follow-up showed stable results, significant improvement in cervical mobility, and high family satisfaction regarding the aesthetic outcome.

Discussion

The surgical correction of *pterygium colli* in Turner Syndrome remains a significant technical challenge due to cervical anatomical complexity and the high risk of recurrence inherent to pediatric growth. Although the literature describes variations such as the five-flap Z-plasty and the use of tissue expanders, the multiple Z-plasty technique employed in this case stands out for its versatility in providing skin extension with favorable aesthetic results, lower morbidity, and a shorter treatment time for a clinically delicate patient. However, the management of pediatric TS patients must transcend isolated technical planning, requiring strict integration with multidisciplinary care [2,4,5].

The partial recurrence of the otoplasty observed in this case, initially performed at age 4, raises relevant questions regarding tissue biomechanics in TS. Literature suggests that cartilage in TS patients may possess distinct structural properties influenced by chronic peripheral lymphedema—which alters microcirculation and connective tissue architecture from the fetal period—and the characteristic hypoestrogenism, which impacts fibrocartilaginous density and resistance. Furthermore, the therapeutic use of growth hormone (GH), while essential for height development, promotes continuous chondral remodeling that may confer a more resilient tissue "memory," increasing the technical challenge of maintaining symmetry after early otoplasties. These findings reinforce the need for more robust cartilage fixation techniques and surgical planning that accounts for structural instability during rapid growth phases.

Endocrine follow-up plays a central role, especially regarding GH use and estrogen replacement. The timely initiation of these therapies must be articulated with the reconstructive schedule to avoid conflicts between growth spurts and critical healing periods. It is imperative to note that while GH aids height development, it does not correct structural deformities, highlighting the need for continuous plastic surgery surveillance during skeletal maturation.

Medial epicanthus is one of the most frequent ocular manifestations in TS, present in up to 46% of patients; its correction is essential for facial harmony and unobstructed peripheral vision. In this case, rotation flaps were chosen for bilateral medial epicanthus correction. This technique allowed for precise tissue repositioning and the release of scar tension in the inner canthus. Unlike purely decisional approaches, rotation flaps offer greater predictability in scar positioning, resulting in satisfactory facial symmetry and a more natural postoperative palpebral contour. This technical choice is particularly advantageous in longitudinal pediatric management, as it minimizes the risk of secondary retractions during facial growth, significantly contributing to improved self-image.

Furthermore, perioperative safety is conditioned by cardiometabolic vigilance, as TS is associated with a mortality risk up to three times higher than the general population, predominantly due to cardiovascular causes. Even in asymptomatic patients, screening via echocardiography and advanced imaging (MRI/CT) is essential to mitigate risks during general anesthesia, given the incidence of aortic dissection at early ages [6-8].

In this context, the sequential and longitudinal correction presented allowed not only for cervical and facial harmony but also for the strengthening of the patient's self- image. In a scenario where adult women with TS frequently report emotional vulnerability and gaps in transitional care, early intervention and longitudinal follow-up become fundamental tools to improve quality of life and ensure consistent long-term clinical outcomes [7,9,10].

Conclusion

The surgical management of Turner Syndrome in pediatric patients requires sequential planning that considers both the technical complexity of the deformities and the dynamism of osteochondral growth. The use of multiple cervical Z-plasties and medial epicanthus correction via rotation flaps proved to be safe and effective strategies, providing facial and cervical harmony with lower morbidity in a delicate clinical scenario.

The 5-year longitudinal follow-up demonstrates that aesthetic and functional success depends on strict integration with endocrine therapy and cardiological surveillance, ensuring that reconstructive interventions occur at optimal developmental stages. Furthermore, understanding the biomechanical properties of cartilage and soft tissues in TS is fundamental to mitigating recurrence risks and optimizing long-term results.

Ultimately, this report reinforces that early and progressive intervention is essential to mitigate psychosocial stigmas and strengthen self-image. Continuity of care and the consolidation of multidisciplinary protocols are the pillars for ensuring consistent clinical outcomes and improving the quality of life for these patients from childhood into adulthood.

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