

Amniotic Band Syndrome in Adult Combined with Persistent Depressive Disorder

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Summary: Congenital amniotic band syndrome (ABS) is an anomaly with no proven etiology occurring in 0.7 per 10,000 live births. This defect mostly concerns the extremities and is often accompanied by other developmental anomalies. There are many methods of treatment for this type of defect, such as simple excision and suturing, local V-Y plasty, Z-plasty, multiple Z “plasties” or multiple W plasties, plasty with deepithelized or non-deepithelized rectangular lobes, and rigotomies complemented with lipofilling. The literature most often describes cases of treated children. There are no case reports of ABS treatment in adults. However, failure to undertake such treatment in childhood may result in serious mental dysfunction. We present the case of a 39-year-old woman with congenital ABS, in whom failure to provide proper treatment in childhood resulted in persistent depressive disorder development. The applied treatment, consisting of multiple Z plasties, liposuction, and fat grafting, resulted in improved appearance of her lower extremity, as well as the cessation of mental symptoms. (*Plast Reconstr Surg Glob Open* 2021;9:e3594; doi: [10.1097/GOX.0000000000003594](https://doi.org/10.1097/GOX.0000000000003594); Published online 26 May 2021.)

Congenital amniotic band syndrome (ABS) is an anomaly with no proven etiology occurring in 0.7 per 10,000 live births.¹ This defect mostly concerns the extremities and is often accompanied by other developmental anomalies.² There are many methods of treatment for this type of defect.³⁻⁵

The literature most often describes cases of treated children. There are no case reports of ABS treatment in adults. However, failure to undertake such treatments in childhood may result in serious mental dysfunction.⁶⁻⁹ Here, we present a rare case of ABS in an adult patient and describe our management approach.

CASE REPORT

A 39-year-old female patient was referred by an orthopedic surgeon for treatment in our hospital due to circular constriction between the middle and lower third of the left leg (Fig. 1). In addition, the patient had other abnormalities of the left hand and right and left feet (Figs. 2, 3). Failure to apply proper treatment in childhood was a source

of significant psychological discomfort for the patient, manifesting as depressed mood, low self-esteem, and feelings of hopelessness for more than 2 years. According to her psychiatrist, the patient met the diagnosis of persistent depressive disorder (300.4, F34.1), as evidenced by her chronic anhedonia and avoiding going to summer holidays to avoid displaying her physical abnormalities. This triggered impairment in her daily functioning.

First, a rigotomy¹⁰ combined with a fat grafting¹¹ was performed. No significant improvement was achieved multiple Z “plasties” of the half posterior constriction part were performed. A partial improvement of the appearance and a definite improvement of the patient’s mental condition were achieved. Multiple Z plasties of the remaining anterior part of the constriction were performed after 5 months. There was another improvement in the appearance of the leg. The final stage of treatment was the liposuction of the calf. Follow-up after 5 years showed natural appearance of the left leg; the scar after multiple Z plasties was smoothed and faded (Fig. 4). The treatment has significantly improved patient’s self-esteem and mood. After the surgery, patient displayed confidence in wearing clothes that accentuated her appearance and, for the first time, she did not avoid vacation

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Fig. 1. Appearance before the treatment. Constriction between the middle and lower third of the left leg, with excessive adipose tissue development above and below the fibrous ring.



Fig. 3. Total syndactyly of the toes in the right foot, syndactyly of toes 1 and 2 of the left foot, and absence of toe 4 of the left foot.



Fig. 2. No distal phalanx of the 3rd finger in her left hand, with circular constriction in the distal part of the 4th finger in her left hand.

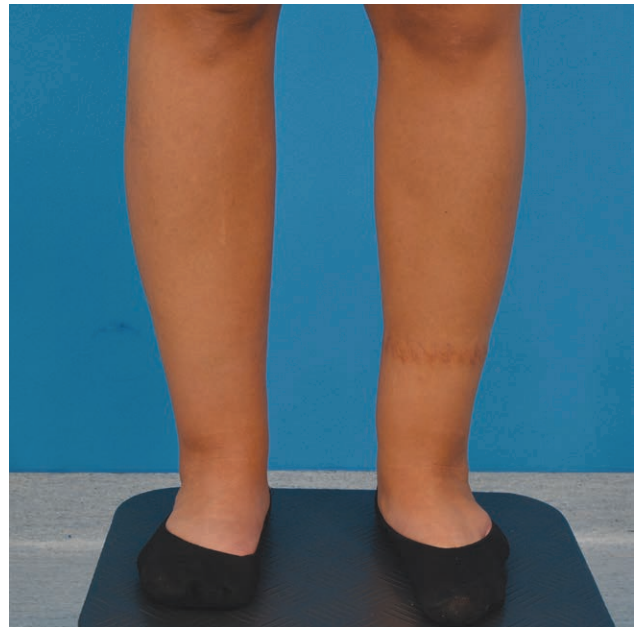


Fig. 4. Appearance 5 years after the treatment. Visible correction of the left leg outline and fainting and smoothing of the scar after multiple Z plasties.

that involved going on the beach. The patient's mental status was subjectively determined without a follow-up with her psychiatrist.

DISCUSSION

Congenital constriction ring syndrome, also known as amniotic disruption sequence or ABS, has been referred

to in the literature using 34 different names.¹² The syndrome was first described by Montgomery in 1832.¹³⁻¹⁶

ABS comprises a group of anomalies that correspond to 1%–2% of congenital malformations in the general population.¹² ABS prevalence is 1 in 1200 to 15,000 live births.^{12-14,17-20} The male-to-female ratio is 1:1, and it occurs most commonly in the Afro-Caribbean population.^{12,15} The syndrome affects both upper and lower extremities at

Table 1. Classification of ABS according to Patterson

Type I—simple ring constrictions
Type II—ring constrictions accompanied by deformity of the distal part with or without lymphedema
Type III—ring constrictions accompanied by fusion of distal parts ranging from mild to severe acrosyndactyly
Type IV—intrauterine amputations at any level

a ratio of 85% to 15%, respectively,^{16,21} most often involving fingers.

ABS is accompanied by other upper and lower extremity defects such as clubfoot, leg length discrepancies and bone abnormalities, oligodactyly, syndactyly, polydactyly, and micrognathia. Craniofacial defects such as cleft lip or palate, plagiocephaly, cranial changes to acrania, and other anomalies such as renal cysts and intestinal obstructions are also described.^{13–16,20–23}

The reported causes of ABS include genetic conditions, the course of pregnancy with concomitant diseases such as diabetes and hypertension, infections, stimulant, and medicine use. However, the exact pathogenesis of this disease remains unknown.^{13,15,23}

There are 3 theories of ABS pathogenesis. (See appendix, Supplemental Digital Content 1, which displays 3 theories of ABS pathogenesis. <http://links.lww.com/PRSGO/B663>.)

The classification proposed by Patterson in 1961¹ distinguishes 4 types of this anomaly (Table 1). The most common type is type III, including both atrophy and lymphedema, related to 75%–88.5% of cases.²¹

The treatment period for this type of disease depends on the severity of the defect and the diagnostic capabilities. If the syndrome is diagnosed during the prenatal period, the surgery is performed possibly still in the womb.^{24–26} If a constriction is noticed immediately after birth and poses a risk of amputation of the limb part or other ischemic defect, the procedure should be performed immediately. However, if the amniotic constriction does not pose an amputation threat, but may have a negative effect on the normal development of the extremity (type II), the procedure is performed at the age of 3–6 months. When dealing with type I constriction, it is recommended to perform surgery before the child is 2 years old, due to psychological reasons^{18,21} (Table 1).

Among the treatment methods, the most frequently described include simple excision and suturing, local V-Y plasty, Z-plasty, multiple Z plasties or multiple W plasties.^{14,16,18,21,22} In 2006, Mutaf and Sunay used plasty with deepithelized or nondeepithelized rectangular lobes.³ Khouri introduced the concept of percutaneous aponeurotomy and rigotomies complemented with lipofilling.¹¹

In the presented case, no improvement was achieved after the fat grafting combined with rigotomy; therefore, multiple Z plasties were applied. A satisfactory aesthetic effect was achieved, providing stable improvement in the appearance of the leg, confirmed in follow-up after 5 years.

ABS not only causes a change in the body's appearance, but can also effect mood. In the described case, the failure to perform the corrective procedure in childhood caused the patient to develop a psychiatric illness in the form of

persistent depressive disorder. This caused her to have significant impairment and lowered her life quality. If the procedure had been carried out at least during adolescence, when one is still defining own body image, perhaps her psychiatric symptoms might have been avoided altogether.^{9,10}

CONCLUSIONS

ABS is a congenital defect that causes both upper and lower limb deformities, but can also result in development of psychiatric symptoms. The use of multiple Z plasties combined with liposuction and fat grafting not only significantly improved the appearance of the treated extremity, but also improved the patient's mental state both objectively and subjectively. All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

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