Case Report
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Amniotic Band Syndrome: A Report of Two Cases by a Primary Care Pediatrician

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Abstract

Objective – Amniotic Band Syndrome is a rare birth defect caused by strands of the amniotic sac that tangle around the baby's body, causing injury. Amniotic Band Syndrome is not seen very often, but should be considered in every newborn with congenital anomalies. It can be diagnosed prenatally by ultrasound and is seen after birth. A team of specialists, a plastic surgeon, an orthopedic surgeon, a physiatrist, a psychologist and others should be included in the treatment and follow-up of children with Amniotic Band Syndrome, with an individual approach to every case. **Case Report** – The first patient, who was six years old, had malformations of the left hand, rudimentary fingers II and IV, missing finger III with circulatory ring, and clinodactyly of finger V. The second patient, eight months old, had the following malformations: syndactyly and circulatory ring on fingers II, III and IV of the left hand, and lymphedema; circulatory ring and lymphedema of the left lower leg, visible proximal parts of all five fingers; on the right foot syndactyly of toes I, II and III, a circular groove on toe IV, and lymphedema. **Conclusion** – Our cases have unique clinical presentations and therefore required reports. In such cases, the primary care pediatrician brings together a multidisciplinary team and provides further follow-up through rehabilitation and psychological treatment.

Key Words: Amniotic Deformity ▪ Streeter's Dysplasia ▪ Constriction Rings.

Introduction

Amniotic Band Syndrome (ABS) (Streeter's dysplasia, Congenital Constriction Band Syndrome, Amniotic Band Sequence, Ring Constriction Syndrome, Amniotic Deformity) is a rare birth defect caused by strands of the amniotic sac that become entangled around the baby's body, causing injury, and they are responsible for the abnormal development of fetal parts. Amniotic Band Syndrome manifests clinically as constriction rings which lead to lymphedema of the digits, other complex, multiple congenital malformations and defects of multiple body parts, some incompatible with life (1). It affects both sexes equally, with an incidence of 1 in 1,200 to 15,000 live births (2), 1 in 70 stillbirths (3) and 178:10,000 spontaneous abortions (1). There is no genetic predisposition. Evidence of familial transmission is extremely rare (4). Three percent of all malformations are physical, and ABS accounts for 1-2% of these physical malformations (5). In the antenatal period, these can be diagnosed by ultrasonography, and after birth by clinical examination, ultrasonography, X-ray, or magnetic resonance. The treatment involves a multidisciplinary approach, primarily surgical, with an individual approach to every single case.

The aim of this paper is to present new cases of patients with ABS, new examples with different combinations of deformity, considered to be part of the Amniotic Band Syndrome spectrum.

A Case report

Case 1. A boy aged 6 years, the first child from the second pregnancy (1 miscarriage) of a healthy

mother aged 26 years. There was no history of maternal alcohol, tobacco or drug intake, radiation exposure, trauma, amniocentesis, chorionic villous sampling, or any history of infection during pregnancy. In the family, there was no prenatal chromosomopathy, consanguinity or malformations. The pregnancy was regularly controlled, ultrasound examination did not show any fetal malformations. The mother was taking progesterone from 5 gestational weeks due to vaginal bleeding. During the last three weeks of pregnancy, she had swelling of the legs, and five days before the birth she also had hypertension and received methyldopa. Delivery was vaginal and occurred in the 39th, 4/7, gestational week. Birth weight was 4550 g (>90th percentile), birth length 56 cm (>90th percentile), head circumference 35 cm (50th percentile), Apgar score 10/10. The patient's developmental milestones were normal. The examination showed the following malformations: left hand: hypotrophy, thumb smaller, rudimentary fingers II and IV with proximal phalanxpresent, missing finger III with a round stump of 5 mm and circulatory ring, and clinodactyly of finger V (Fig. 1 (A, B). Besides, there were a few minor malformations present: asymmetricalears, short neck, sandal furrow on the feet on both sides; overall hypotonia and obesity. All examinations performed after birth (ultrasound of the central nervous system and abdominal organs, echocardiography, and ophthalmoscopic examination) showed normal anatomy of the examined organs. Karyotyp was normally male (46, XY). The X-ray shows the lack of the medial and distal phalanges of fingers II and IV, and the lack of the entire finger III (Fig. 1 (C).

The dysmorphological signs described: asymmetrical ears, short neck, sandal furrow on the feet on both sides, cannot be reliably correlated with ABS. A plastic surgeon was consulted and plastic correction is planned in adulthood when the child stops growing. He attends physical rehabilitation treatment regularly, as well as receiving psychological support.

Case 2. Male infant, the third child from the fourth pregnancy (one artificial abortion) of a healthy mother, aged 33 years. There was no history of maternal alcohol, tobacco or drug intake, radiation exposure, trauma, amniocentesis, chorionic villous sampling, or any history of infection during pregnancy. In the family, there was no prenatal chromosomopathy, consanguinity or malformations. Pregnancy was controlled regularly, ultrasound examination did not show any fetal malformations. Delivery was vaginal and occurred in the 38th, 2/7, gestational week. Birth weight was 4130 g (75 percentile), birth length 50 cm (50 percentile), head circumference 36 cm (50 percentile), Apgar score 9/10. The examination showed the following malformations: on the left hand syndactyly of fingers I, II, III and IV, circulatory ring on fingers II, III and IV of the left hand and lymphedema; circular ring on the left lower leg, lymphedema of the left lower leg, visible proximal parts of all five fingers; on the right foot syndactyly of toes I, II and III, circular

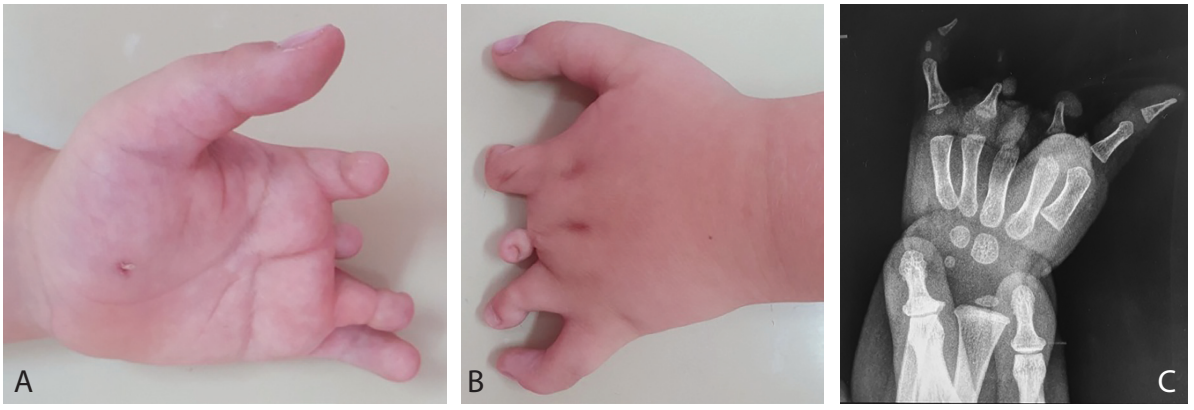

Fig. 1. *First patient, left hand, palmar side (A); First patient, left hand, dorsal side (B); First patient, X-ray of left hand (C).*

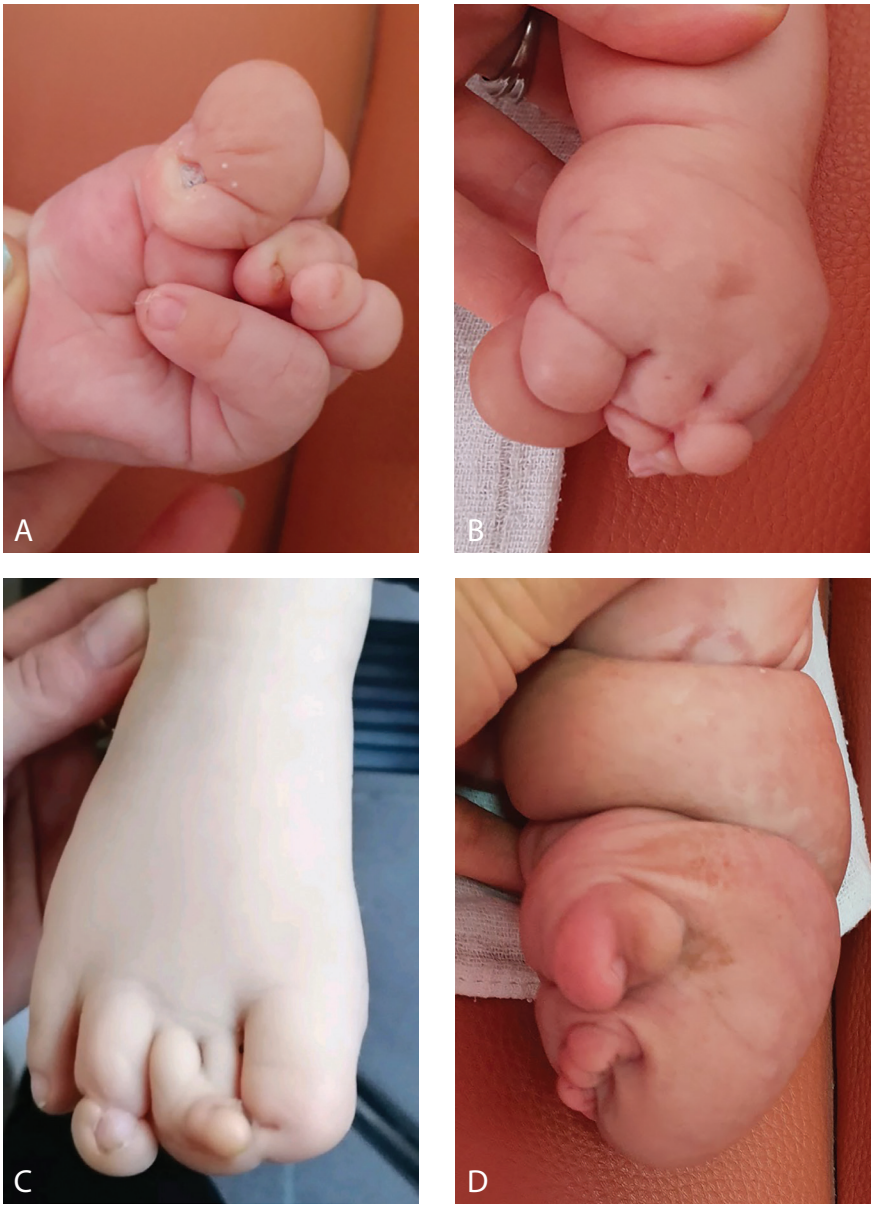

Fig. 2. *Second patient, left hand, palmar side (A); Second patient, left hand, dorsal side (B); Second patient right foot (C); Second patient, left foot (C).*

groove on toe IV, and lymphedema (Fig. 2. A, B, C, D). Reducible umbilical hernia present. Cardiology and echocardiography: Foramen ovale apertum of minimal L-R shunt, Chorda aberans ventriculi sinistri. Ultrasound of the central nervous system and abdominal organs, ophthalmoscopic examination showed normal anatomy of the organs examined. Karyotype was normally male (46, XY). Other symptoms described: reducible umbilical hernia, Foramen ovale apertum and Chorda aberans ventriculi sinistri cannot be reliably associated with ABS. A plastic surgeon, orthopedist and physiatrist were consulted, and serial plastic corrections are underway, with physical, psychological and orthopedic support.

Discussion

These deformities cannot be explained by a single pathogenesis. The etiology of ABS is still unknown. A combination of amniotic disruption, vascular disruption, embryonic dysplasia and genetic disorganization can lead to the development of the syndrome (6). The most accepted theory is amniotic disruption or extrinsic theory, which was proposed by Torpin in 1965. According to this theory, amniotic rupture is followed by extrusion of parts of the fetus into the chorionic cavity, and compression, with the fetal limbs or other body parts becoming entangled in the rolled remnants of the amnion. The type of anomalies depends on the timing of the amniotic rupture during pregnancy, as well as the location and degree of compression of the underlying structures (6). Risk factors, such as maternal age, trauma, infections, prematurity, amniocentesis, chorionic villus sampling, and drugs, have been

considered. The mothers of the children we have shown are 26 and 33 years old. A connection has been found between Amniotic Band Syndrome and the mother's age - it is more common in primiparas under the age of 25 (1). Trauma, infections, radiation, prematurity, amniocentesis, and alcohol and drug use were denied in both pregnancies. Both cases shown had no chromosomopathy, consanguinity or malformations in the family. There are many different combinations of anomalies and deformity, depending on the location, and there are no two identical ABS cases. Due to this unique specificity, we have presented these two cases that were found at our clinic.

Patients with Amniotic Band Syndrome have very polymorphic physical findings: craniofacial anomalies, various facial clefts, cleft lip and palate, orbital anomalies, central nervous system malformations and calvaria defect (7), truncal defects, cardiac abnormalities, congenital diaphragmatic hernia, renal agenesis, internal genital malformations, anal atresia (8), and limb defects: constriction rings, lymphedema of the digits, shortening of the limbs or intrauterine limb amputation, amputation of the digits (most often fingers II, III and IV), syndactyly, hypoplasia of the digits, peripheral nerve palsy, high death rate, and incompatibility with life (9, 2). Patterson's diagnostic criteria for congenital ring constrictions: 1. Simple ring constriction; 2. Ring constrictions accompanied by deformity of the distal part, with or without lymphedema; 3. Ring constrictions accompanied by fusion of distal parts ranging from fenestrated or terminal syndactyly to "exogenous" syndactyly; 4. Intrauterine amputations. One or more criteria must be present (10).

Amniotic Band Syndrome can be diagnosed prenatally by three-dimensional and four-dimensional ultrasound examination, sometime as early as at 12 gestational weeks, if constriction, amputation, distal limb edema or deformation of major anatomic structures are present, but most commonly in the second trimester of gestation during routine ultrasound examinations. In a very few cases a strand of amniotic fibrous tissue can be seen attached to tissues and restricting the free movement of the fetus in-utero (11). A histopathological examination may reveal the absence of the amniotic membrane on the fetal surface of the chorionic sac, including the placenta (12). Sometimes, ABS can be diagnosed by fetal MRI (3). Clinical examination is the basis of postnatal diagnostics of Amniotic Band Syndrome, with the use of additional examinations by ultrasound, magnetic resonance or X-ray, in order to find invisible defects of different organs and body parts (13). Differential diagnosis of amniotic bands may be that they are from uterus and placental anomalies or limb amputations due to other causes of congenital absence of limbs. There are numerous syndromes with associated congenital limb anomalies (11). The child's chromosomal karyotype analysis is needed in order to exclude a mistaken diagnosis. In our patients, the diagnosis was made postnatally: by clinical examination, ultrasound, and X-ray, and genetic processing was also performed.

ABS treatment can be prenatal or after birth. In cases when amniotic banding is constricting the umbilical cord or cutting off the blood supply to a baby's limb, a surgeon can attempt fetoscopic amniotic band resection to surgically remove the bands, guided by ultrasound imagery, a laser or other surgical instrument before the baby is born (11). The treatment after birth involves a multidisciplinary approachinvolving a plastic surgeon, orthopedic surgeon, orthodontist, ophthalmologist, and neurosurgeon, with an individual approach to every single case (14). This is also useful in properly advising pregnancy termination in fetal anomalies incompatible with life (15).

Conclusion

Amniotic Band Syndrome is a rare congenital malformation with a wide spectrum of clinical presentations that are unique to each patient. Our cases have unique clinical presentations, none of which has been previously described in the literature and therefore they require a report. For the future, we recommend frequent expert US examinations (four-dimensional ultrasound) in order to diagnose

the syndrome in good time, and possibly perform prenatal therapy. The primary care pediatrician brings together a multidisciplinary team, and provides further follow-up through rehabilitation and psychological treatment. Prevention centers that specialize in surgery to treat amniotic band syndrome often provide support networks to the child and parents. There are several active Facebook groups for families of children with amniotic band syndrome and birth defects in general. Sharing knowledge and stories can help parents to feel connected as well as more empowered during an uncertain time.

Conflict of Interest: The authors declare that they have no conflict of interest.

Declaration of Patient Consent: The authors certify that they have obtained all appropriate patient consent forms. In the forms, the patients gave their consent for their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity. We received no financial support or sponsorship.

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