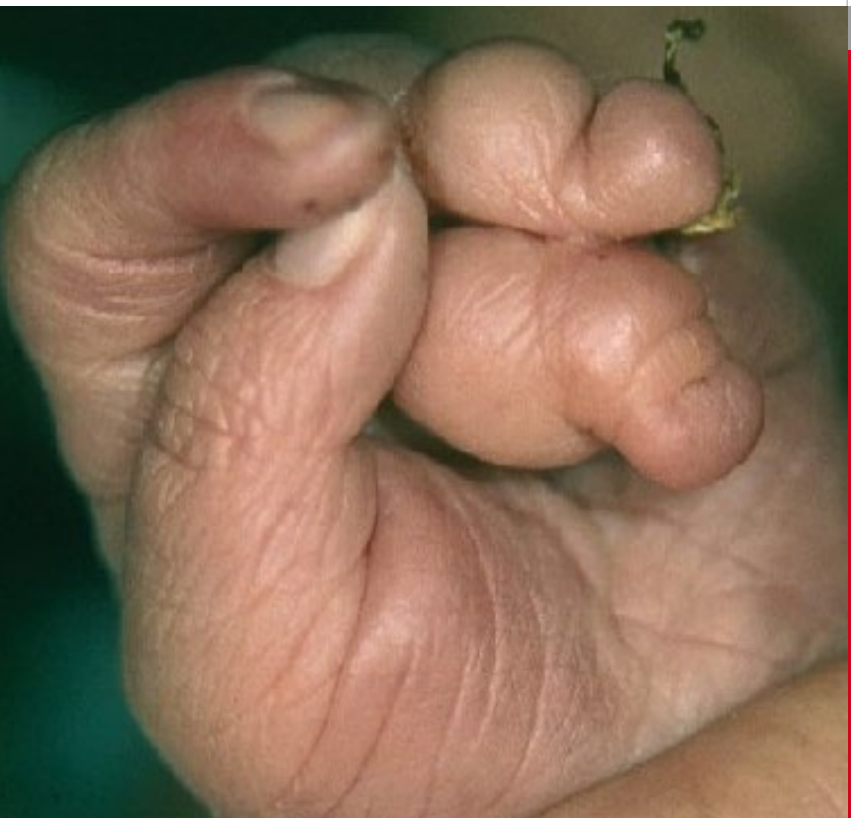
SWISS SOCIETY OF NEONATOLOGY

Amniotic band syndrome



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This male infant was born at a gestational age of 35 2/7 weeks. Pregnancy had been complicated by PROM at 23 weeks of gestation. The mother's medical and obstretical history were otherwise unremarkable. There was no family history of any congenital anomalies. After delivery, the infant showed signs of respiratory distress secondary to a pneumothorax which resolved quickly after drainage. Birth weight was 2730 g, length 44 cm, and head circumference 32 cm.

On admission, the baby was pink in room air. The amniotic membrane was fused with digits III to V of the left hand with amputation of digit III and incomplete syndactylia of digit IV and V (Fig. 1, 4), digits III to V of the right hand with multiple ring constrictions. Almost all limbs showed stunted growth of fingers and toes with dystrophic nails. Incomplete agenesis of digits II to V of the left foot was also found (Fig. 2, 3, 5). Growth of the right foot was also stunted and, in addition, showed a club foot deformity.

A diagnosis of anmniotic band syndrome (ABS) was made. Synonymous expressions for ABS are: ADAM complex (Amniotic Deformity, Adhesions and Mutilations), amniotic band disruption complex or sequence, amniotic bands and sheets, annular constriction bands, congenital ring constriction, congenital transverse defects, constriction band syndrome, intrauterine amputation, limb-body wall complex, Streeter's dysplasia, TEARS (The Early Amnion Rupture Spectrum).

Fig. 1

Left hand with constriction bands, and fusion of digits II-V by amniotic membrane.

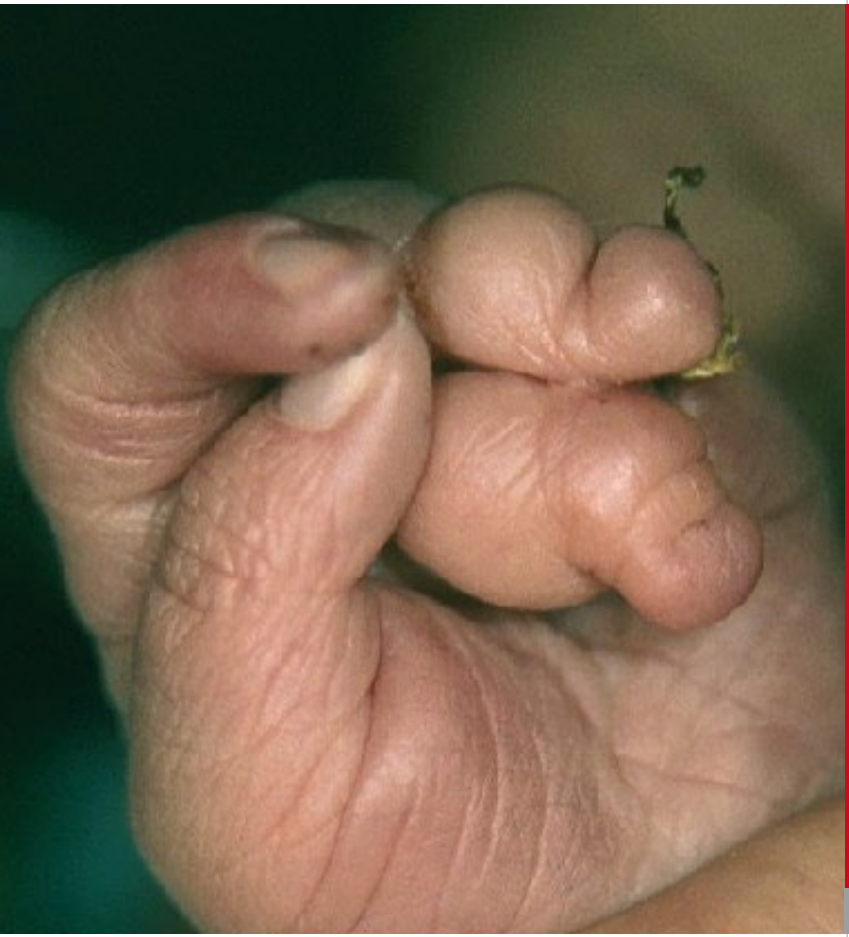

Fig. 2

Left hand with constriction bands, dystrophic nails and fusion of digits II-V by amniotic membrane.

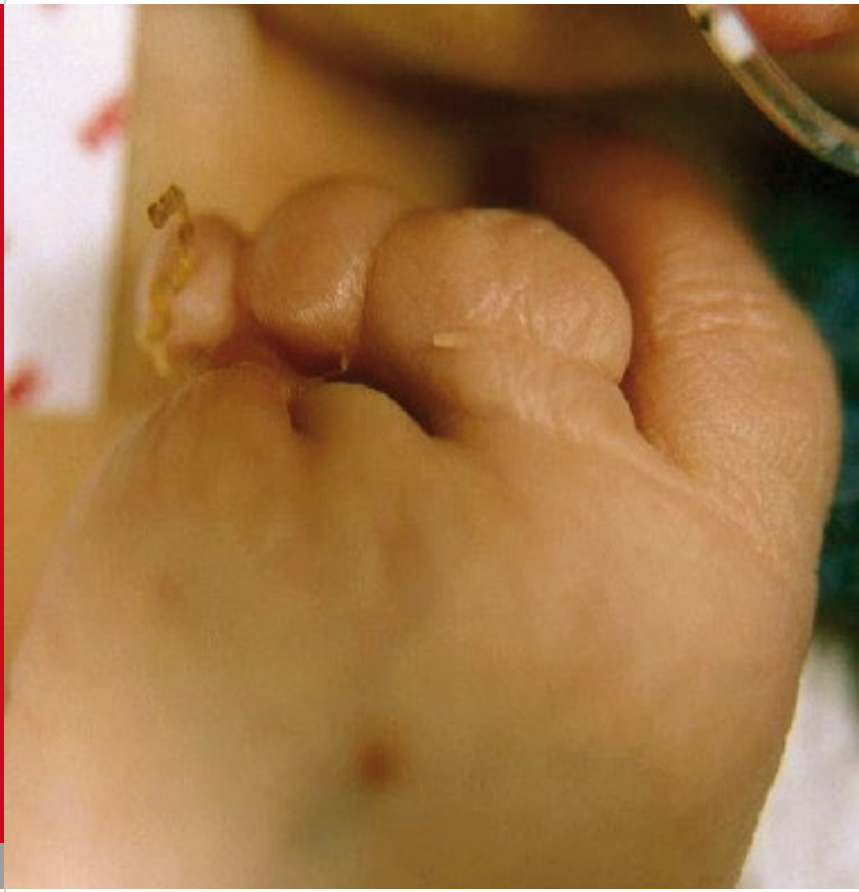

Fig. 3

Left hand with constriction bands, fusion of digits II-V by amniotic membrane.

Fig. 4

X-ray of the left hand.

Fig. 5

Right foot with stunted growth of all digits, dystrophic nails, constriction bands and partial syndactyly of digits II to V.

Fig. 6

Right foot with stunted growth of all digits, dystrophic nails, constriction bands and partial syndactyly of digits II to V.

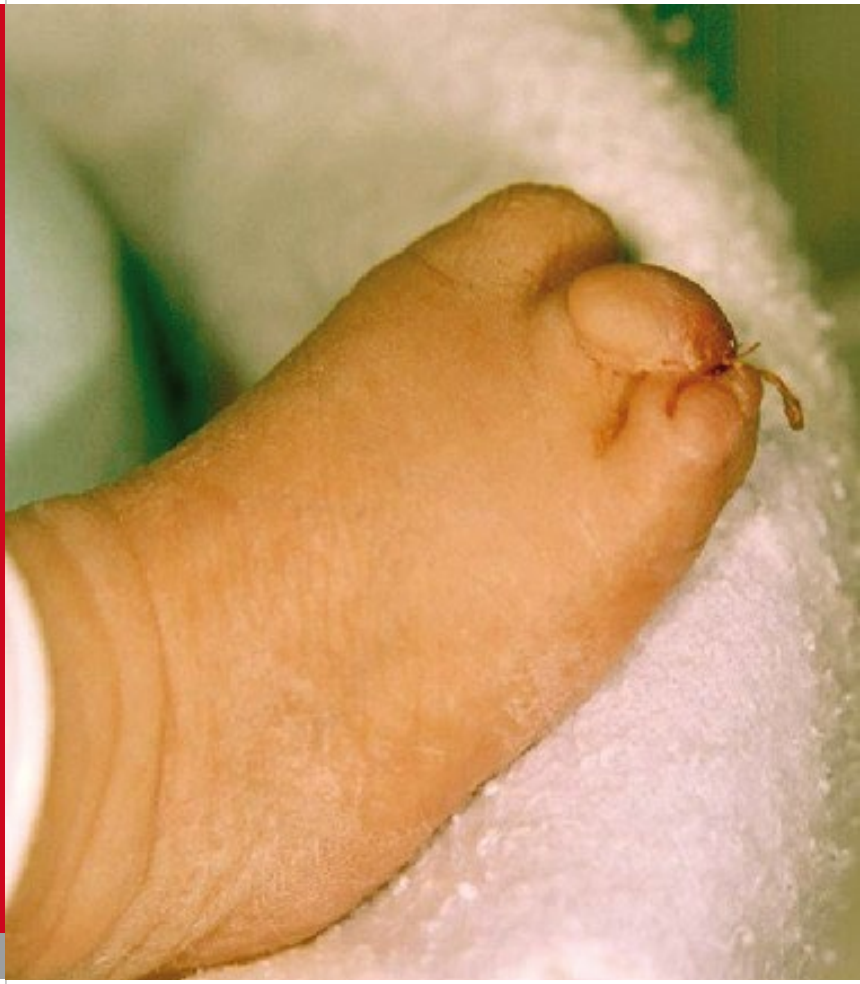

Fig. 7

Right foot with stunted growth of all digits, dystrophic nails, constriction bands and partial syndactyly of digits II to V.

Fig. 8

X-ray of right foot.

Amniotic band syndrome (ABS) is believed to be caused by entrapment of fetal parts (usually a limb or digits) in fibrous amniotic bands while in utero. ABS is not a genetic disease. It is extremely unlikely that ABS will affect a fetus in a future pregnancy. To date, no prenatal factors have been associated with ABS. The etiology is still obscure. The commonly accepted view is that ABS occurs when the inner membrane (amnion) ruptures without injury to the outer membrane (chorion). This exposes the baby to fibrous sticky tissue bands of the placenta which can float in the amniotic fluid and can entangle the baby. In some cases a complete „natural" amputation of a digit(s) or limb(s) may occur before birth or the digit(s) or limbs may be necrotic and require surgical amputation following birth.

There are several features that are relatively consistant in ABS. Distal ring constrictions, limb deformities, and intrauterine amputations are the most common findings. Other abnormalities found with ABS are webbed fingers and toes, progressive lymphedema, clubfoot, clubhands, stunted growth of fingers and toes and limb length discrepancy, cleft lip and palate. Less common are pseudarthrosis, metatarsus adductus, peripheral nerve palsy, dystrophic nails, postnatal gangrene, dislocated hip, visceral body wall malformations, eccentric craniofacial synostosis defects and skin-tubed pedicles. Finally, fetal death associated with amniotic band strangulation of the umbilical cord has also been reported. Seeing that no two cases are

exactly alike, only some of the above features will be present in each individual case.

Treatment is symptomatic and must be individualized. In utero surgery has been performed to free limbs from amniotic bands when amputation seemed imminent. An interdisciplinary approach (plastic, hand, orthopedic surgeons) is often required.

REFERENCES

1. Fiedler JM, Phelan JP. The amniotic band syndrome in monozygotic twins. Am J Obstet Gynecol 1983;46:864-865

2. Keller H, Neuhauser G, Durkin-Stamm MV, Kaveggia EG, Schaaff A, Sitzmann F. ‚ADAM complex' (amniotic deformity, adhesions, mutilations)--a pattern of craniofacial and limb defects. Am J Med Genet 1978;2:81-98 *(Abstract)*

3. Lubinsky M, Sujansky E, Sanger W, Salyards P, Severn C. Familial amniotic bands. Am J Med Genet 1983;14:81-87 *(Abstract)*

4. Van Allen MI, Siegel-Bartelt J, Dixon J, Zuker RM, Clarke HM, Toi, A. Constriction bands and limb reduction defects in two newborns with fetal ultrasound evidence for vascular disruption. Am J Med Genet 1992;44:598-604 *(Abstract)*

5. Higginbottom MC, Jones KL, Hall BD, Smith DW. The Amniotic band disruption complex: Timing of amniotic rupture and variable spectra of consequent defects. J Pediatr 1979;95:544-549 *(Abstract)*

6. Seeds JW, Cefalo RC, Herbert WNP. Amniotic band syndrome. Am J Obstet Gynecol 1982;144:243-248 *(Abstract)*

7. Foulkes GD, Reinker K. Congenital constriction band syndrome: A seventy-year experience. J Pediatr Orthop 1994;14:242-248 *(Abstract)*

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