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Congenital Constriction Band Syndrome Pathophysiology and Treatment

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The clinical manifestations of 88 children with congenital constriction band syndrome involvement of the hand were reviewed. Seventy-five of these children had evidence of digital or limb amputations, with 235 upper limb amputations and 138 lower limb amputations. In the hand, digital amputations were most common in the index, middle, and ring fingers, whereas in the foot, amputations of the hallux were most often noted. Band indentation was often present at multiple levels. Proximal bands may be associated with neural compression. Syndactyly was invariably associated with a proximal interdigital sinus or cleft and was frequently associated with distal amputation.

Examination of a 27-week gestation stillborn specimen having manifestations of congenital constriction band syndrome demonstrated the intrauterine biologic response to band constriction.

The variable clinical manifestations of congenital constriction band syndrome can best be explained as the response of the growing, embryologically defined limb to intrauterine deformation or band-induced compression and ischemia.

Congenital constriction band syndrome is estimated to occur in approximately one of each 15,000 live births [1]. Numerous names have been suggested for this entity. These include annular groove, amniotic bands, Streeter's dysplasia, and ring constriction syndrome. The term congenital constriction band syndrome is most appropriate in view of current understanding of the pathophysiology of this condition.

While most children's hand anomalies are the result of malformation of the embryonic hand, other abnormalities evident at birth are the result of deformity or of disruption of the developing limb or limb bud. Our observations in a large series of clinical cases as well as those derived from dissection of a stillborn fetal specimen suggest that the manifestations of congenital constriction band syndrome represent both intrauterine deformity and disruption. [2]

MATERIALS AND METHODS

Fetal specimen

Examination of a 27-week gestation stillborn specimen demonstrated variable involvement of congenital constriction band syndrome in all four extremities.

Clinical cases

Clinical review of 88 patients with hand involvement as the result of congenital constriction band syndrome was undertaken at the Shriners Hospital, Chicago Unit. Though

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the injury which provokes this condition occurs *in utero*, the average birth weight of the children in our study group was 6 1/2 pounds (range 3 pounds 1 oz to 9 pounds 11 oz). Children were born at an average gestation of 37 1/2 weeks (range 26 to 44 weeks). Other studies have suggested that prematurity is common in this condition [1]. Most children were the products of otherwise normal uncomplicated pregnancies. None had a family history of congenital constriction band syndrome.

The 88 patients in this study group underwent 304 discrete procedures. Multiple procedures involving up to three extremities were often combined under a single anesthesia.

Treatment

Fibrous bands may be noted encircling the fingers or toes of a newborn child (Figure 1). They may be simply removed or unwound, often untethering fingers from one another.

Treatment of band indentation has consisted of band excision and soft tissue recontouring with multiple Z- or W-plasties which may be combined with debulking of the distal segment (Figures 2, 3). Though some authors [3, 4, 5] advocate routine circumferential band release, the procedures in this series were restricted to 65% of the circumference of the digit. Release of circumferential bands was staged to avoid vascular embarrassment to the distal segment [6]. When bands are extremely tight, profound distal edema is managed by band excision, distal debulking and multiple W-plasties. Tight, deep bands may narrow the underlying bone. Skin and underlying fat may be separately transposed as described by Upton [7] to avoid persistent indentation at the site of flap transposition. Deep fascial constriction is sought and released, particularly beneath deeper proximal bands.

Proximal bands affecting the arm are relatively uncommon, but may be associated with distal neural or lymphatic compromise [4, 8]. The insensate hands that result from neural compromise are particularly vulnerable to infection and contracture [9, 10]. Those limbs with lymphatic inadequacy are vulnerable to repeated cellulitis.

One child in this series underwent 50% release of a very tight mid-humeral band at one week of age. Three months later, the remaining half of the circumference was released and recontoured with debulking W-plasty. The child was referred for treatment at one year of age because of persistent median and ulnar nerve paralysis. The arm was

Figure 1. Newborn with visible bands tethering fingers together.

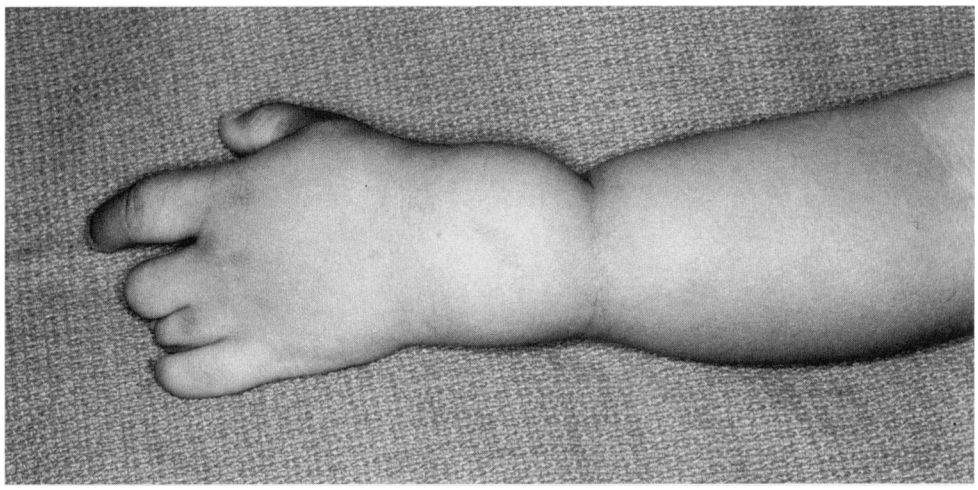

Figure 2. Band constriction has resulted in syndactyly, amputation, and band indentation. (reproduced from [29]).

Figure 3. Forearm Z-plasty, syndactyly release and web space deepening have improved appearance and function. (reproduced from [29]).

re-explored with attention directed to the median and ulnar nerves. Though both nerves were in continuity, each lay beneath a tight, unreleased soft tissue band. This deep fibrous band was excised. Under the operating microscope, neurolysis of both nerves was accomplished. Because of excessive intraneural abnormality within multiple fascicular groups of the median nerve, a segment of the median nerve was excised and replaced with intercalated interposition sural nerve grafts. Histologic examination of the resected tissue demonstrated pseudoneuroma formation proximal to the band with poor nerve quality beneath and distal to the level of compression.

At three years active digital flexion is present. Opponensplasty tendon transfer of extensor *indicis proprius* has helped preposition the thumb. Though it would have been preferable to have released and grafted these nerves at the time of the initial skin release, the prognosis even with early grafting appears guarded in such cases.

Though amputation through the thumb is unusual, when it does occur, hand function may be seriously compromised. The thumb may be lengthened by one of a number of different techniques. Transposition of the index finger, as a modified pollicization on-top-plasty [11] may be appropriate when the index finger has been compromised by distal amputation. Second toe transfer or hallux wrap around flap transfer are useful microvascular alternatives in treating the amputated thumb. The proximal neurovascular and musculotendinous structures of the hand and forearm are normal in congenital constriction band syndrome since these hands initially develop normally. Normal proximal structures are available for anastomosis. This is a more favorable recipient site than the hypoplastic or aplastic hand. In the dysplastic hand, the proximal structures which correspond to the anomalous distal structures are also dysplastic making neurovascular and musculotendinous junctures less than ideal and limiting tendon excursion.

The syndactyly seen in hands affected by congenital constriction band syndrome is usually a distal binding of adjacent digits (acrosyndactyly) almost as though the digits had been lassoed together (Figure 4). The secondary nature of this syndactyly in this condition is confirmed by the consistent finding of an interdigital sinus. This epithelially lined cleft or fenestration penetrates from the volar to dorsal surface (Figure 5). Often the sinus is so small that it is ignored or regarded as only a dimpled indentation in the skin. It is usually possible, however, to demonstrate the sinus by passing a small probe or unfolded paper clip from the palmar side to the dorsum. When an interdigital cleft or fenestration is present with acrosyndactyly, the base of the commissure is usually located more distally than in the normal hand [11, 12].

Syndactyly is released using standard techniques. The sinus is excised to avoid leaving an epithelial rest within one of the released digits. Full thickness skin graft is harvested from the groin crease. Syndactyly release increases functional digital length and

Figure 4. Band constriction has resulted in syndactyly with amputation of distal parts.

enhances digital independence (Figures 6, 7). The authors prefer a dorsally based commissure flap with liberal full thickness skin grafting. When multiple digits are syndactylized, staged release has proven safe and effective. In instances of multiple digital acrosyndactyly, it may be possible to release all of the distally joined finger tips in the first stage. Only nonadjacent commissures are recessed, however, at the time of the first surgical procedure.

When more than two fingers are bound together in an acrosyndactyly pattern, the border digits often force the central digit palmar to the plane of the other fingers [13].

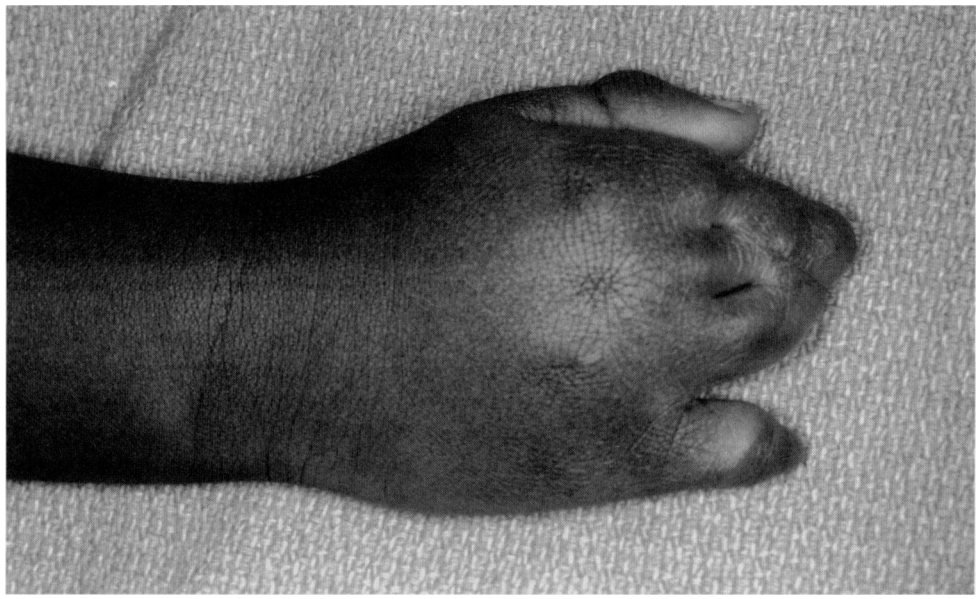

Figure 5. Persistent interdigital sinuses are located further distally than normal web space.

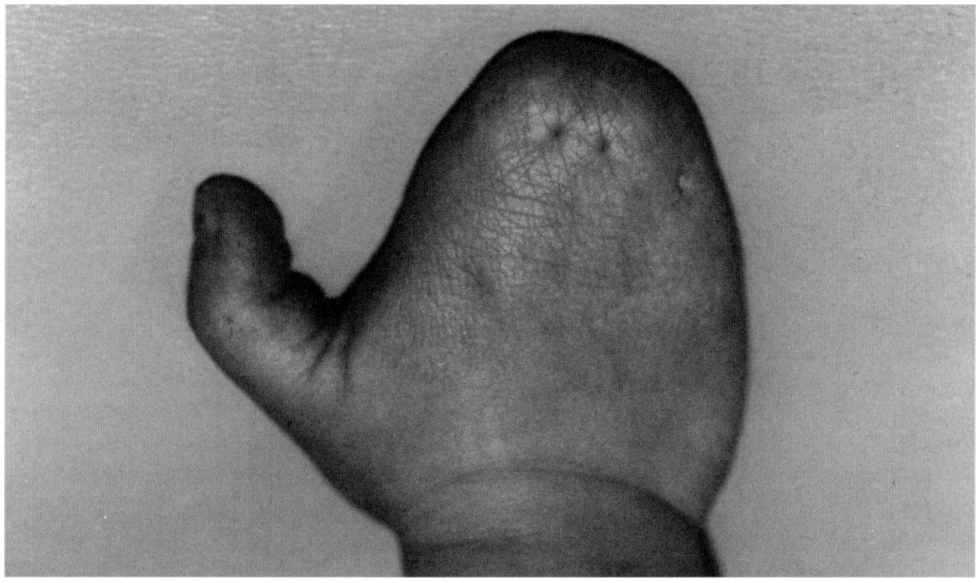

Figure 6. Multi-digit syndactyly secondary to congenital constriction band syndrome.

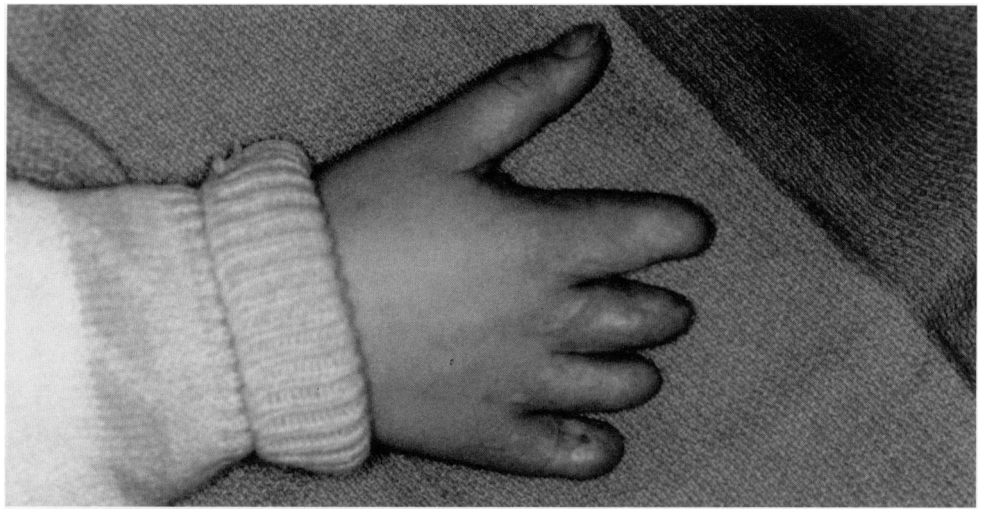

Figure 7. Staged release of hand in Figure 6 has improved appearance and digital independence.

The digits may be drawn together in a tapering conical mass with a mass of grape-like nubbins protruding at odd angles distal to the level of binding. In such hands it may be impossible and unwise to attempt to preserve all distal tissue. It is more sensible to use the portions of these nubbins available to provide good distal soft tissue coverage of the tapered bone ends, deleting floppy unsupported nubbins. When multiple fingers have been joined through the proximal phalanx level, even a modest proximal web space recession will substantially improve digital independence (Figures 2, 3).

RESULTS

Fetal specimen

Morphologic examination revealed fibrous tissue bands encircling the distal third of one leg as well as about the distal portions of many of the upper limb digits (Figure 8).

Band constriction resulted in amputations through joints, as well as through both the metaphysis and the diaphysis of the phalanges. Tapering of the diaphysis was repeatedly noted just proximal to the level of amputation (Figures 9, 10).

Histologic examination revealed an area of progressive tissue destruction distal to the band with cellular autolysis a prominent feature beneath the band. An inflammatory resorptive process was seen both proximal and distal to the band. These changes are consistent with the effects direct pressure and ischemia induced by the constricting band. The foot and limb distal to the band was hypoplastic, suggesting a disruption of intrauterine growth distal to the band.

Clinical cases

Digital band indentation was present in 109 fingers of 81 of our 88 patients (Figure 11). The thumb was the least commonly involved digit, perhaps reflecting its shorter length and its relatively concealed *in utero* posture, adducted into the palm. When the foot was involved, the hallux was the most often affected.

Syndactyly was present in 46 of 88 patients involving 74 different upper and lower

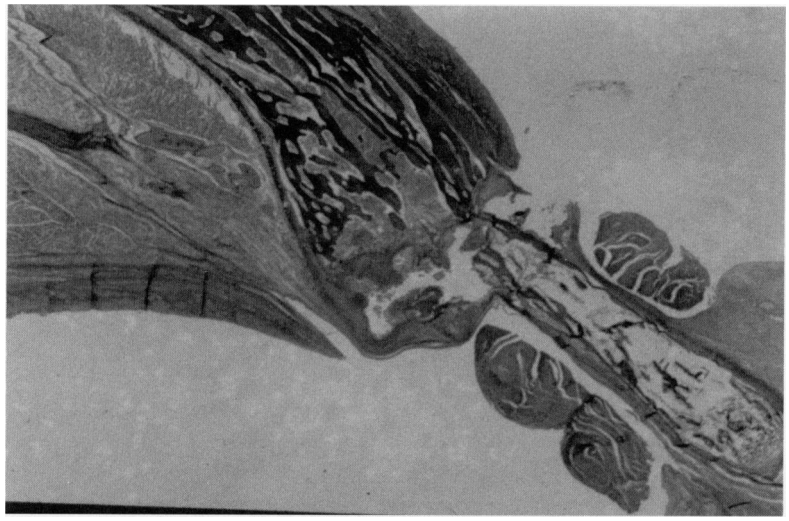

Figure 8. Lateral cross section of fetal tibia demonstrates fibrous band constriction.

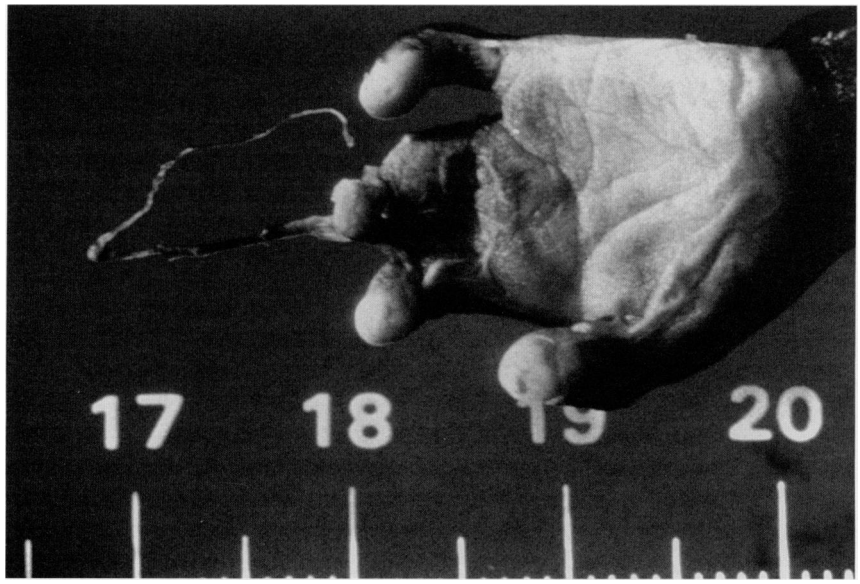

Figure 9. Gross appearance of left hand, with constricting bands encircling ring and middle fingers.

limb sites. In 47 of these sites, amputation was evident just distal to the level of the syndactyly. The middle, ring, and index fingers were most often involved while the little finger was less commonly involved and the thumb involved on only one occasion. Thirty-three of the 46 hand syndactylies were associated with distal amputation, suggesting that a band had pulled adjacent digits together causing distal necrosis and amputation, as well as proximal syndactyly in which the sinus represents the initial interdigital web space (Figure 12).

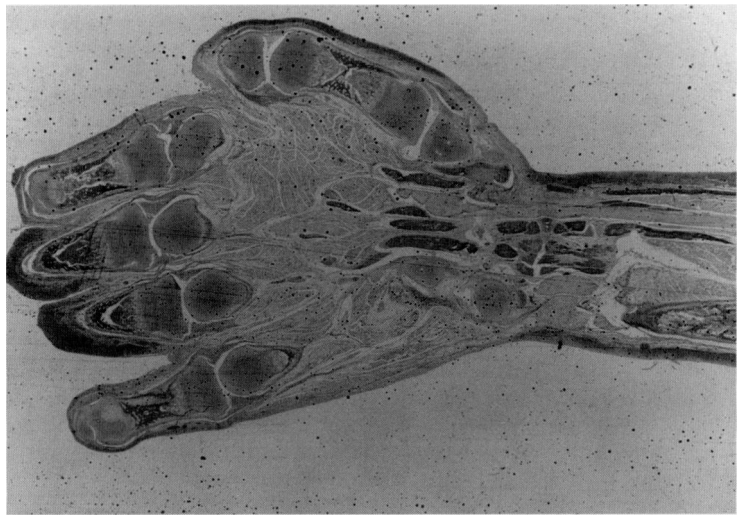

Figure 10. Histologic section through left hand demonstrates tapered diaphyseal amputations of middle and ring fingers as a result of bands evident in Figure 9.

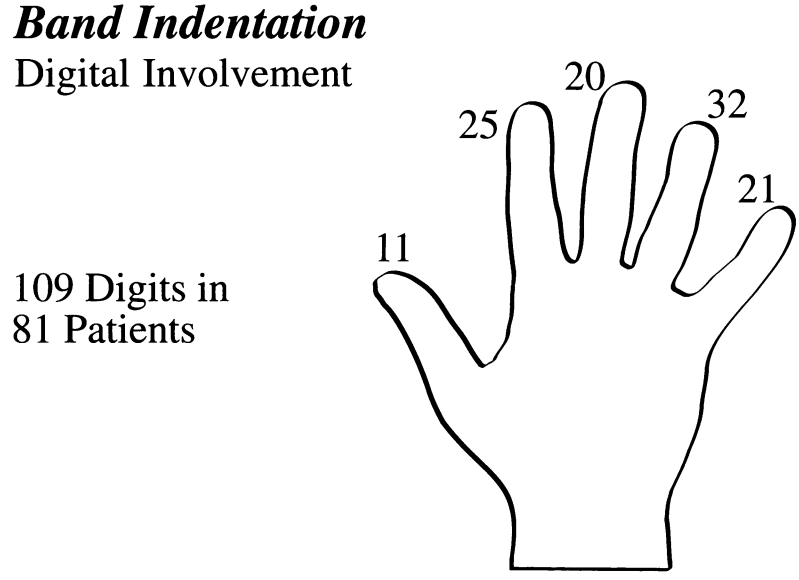

Figure 11. Band constriction location.

Seventy-five of eighty-eight patients had evidence of limb or digit amputation (Figure 13). The fingers were more frequently involved than were the shorter toes. The longer, central three fingers were more commonly amputated than was the little finger, while the thumb, the shortest digit, was the least commonly involved (Figure 12) [14]. Among the toes, the relatively more prominent hallux was the most commonly amputated.

DISCUSSION

The morphology of the various clinical manifestations of congenital constriction band syndrome add further support to the concept of a local compression etiology. When

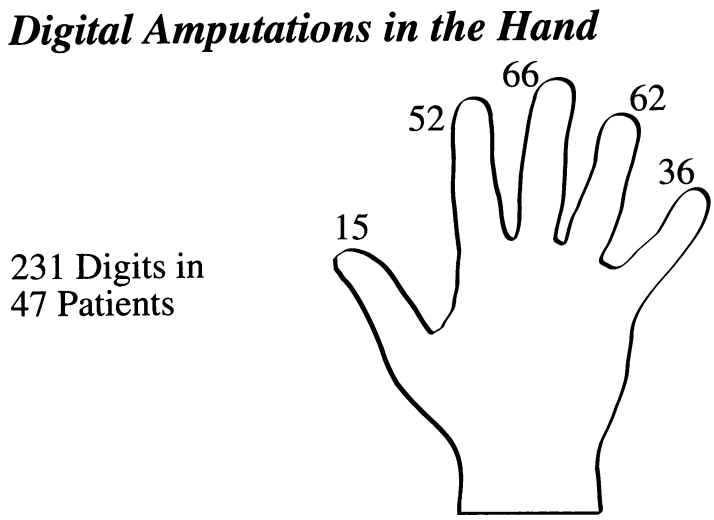

Figure 12. Distributions of amputations in the hand.

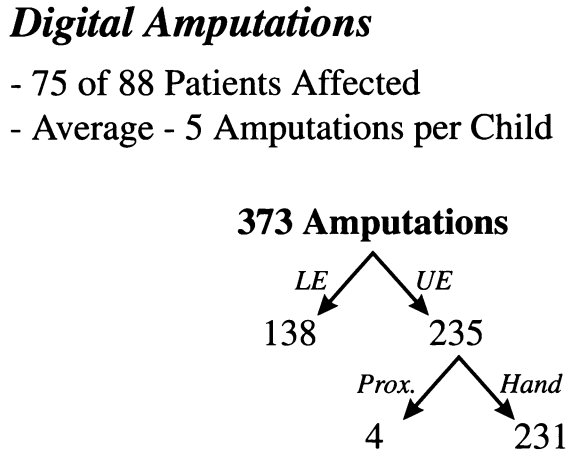

Figure 13. Distribution of upper and lower limb amputations.

bands are superficial, only mild skin indentation occurs. Deeper, tighter bands may produce profound compression with obstruction of both venous and more dramatically lymphatic drainage (Figure 14). Deep compression may result in considerable edema in the digit or limb distal to the band (Figure 15). Deep proximal bands infrequently result in neural compromise. Bands may cause tapering or narrowing of the underlying bone. Even tighter bands may cause amputation of the digit or limb distal to the level of band constriction. Dramatic cases have been reported in which the child was born separate from a

Figure 14. Multiple areas of band indentation are noted in this hand with edema distal to tight bands.

Figure 15. Profound edema in foot and lower leg distal to multilevel constriction.

free amputated piece of distal arm or leg which was delivered separately. These cases give dramatic evidence that amputations can occur *in utero* and, as such, represent a very different entity from failures of formation of parts. When the initially normal digits are pinched together *in utero*, secondary syndactyly results.

The radiographic appearance of the hand and limb is normal proximal to the area of constriction. This is quite different from the hypoplastic, malformed hand in which metacarpal hypoplasia is apparent proximal to the level of digital absence [15, 16].

The etiology of congenital constriction band syndrome condition has been subject of debate for more than 300 years. In 1652, Van Helmont [17] described a child born with a congenital amputation which he believed was the result of the child's mother having viewed a soldier with an amputated arm during the course of her pregnancy.

The description of W.F. Montgomery [18] in 1832 was the first to suggest that these abnormalities were the direct result of intrauterine threads which had become entwined about the developing fetus. Montgomery described finding "complete ligatures about the arms and 2/3 of the leg" in an infant with an intact skin bridge.

Montgomery's explanation was challenged 100 years later by George Linus Streeter [19], the director of embryology at the Carnegie Institute. Streeter believed, wrote, and taught that the etiology of the condition was primarily defective germ plasm. He defended his view point vigorously for more than 35 years. Because of his work many physicians continue to refer to this condition as Streeter's dysplasia.

In 1965, Richard Torpin [20, 21, 22], an obstetrician, challenged Streeter's theory and reintroduced the intrauterine fibrous strand concept of deformity. His comprehensive examinations of placental tissue from involved infants strongly supported the primary role of bands of fetal membrane entwining or entrapping fetal tissues leading to constriction, fusion and or necrosis of distal parts.

In 1975, Kino [23] created a rat model of this condition by performing amniocentesis in pregnant rats 15 days post conception. His work added further evidence to the concept that prenatal environmental factors could induce many of the various manifestations of congenital constriction band syndrome. He further speculated that the injury responsible for congenital constriction band syndrome most likely takes place after major organogenesis, that is after 6 weeks of gestation.

The writings of Higginbottom and co-workers [24] suggests that the manifestations of this syndrome evident in an individual child may be influenced by the timing of the developing limb insult. If the insult occurs early in gestation distal hypoplasia or malformation may be seen, while if the insult occurs later the distal parts are more likely to be of normal size, if present. The abnormalities in the hands are entirely the result of intrauterine deformation.

Tada et al. [9] reported a 27.9%, Patterson [1] a 17.3%, and Moses [25] a 31.1% incidence of club foot in their large clinical series of congenital constriction band syndrome. Twenty-seven (31%) of the 88 children in this series had club foot deformity. In some children this was easily explained by a band in the supramalleolar area [26]. In other children, the club foot was attributed to more proximal bands which could be compromising peroneal nerve function. Other cases have been encountered in which the fingers show the typical manifestations of congenital constriction band syndrome but no discrete lower limb band abnormalities are present to explain the club foot. It has been suggested that one of the precipitating factors for these otherwise unexplained instances of club foot may be oligohydramnios. It has been conjectured the decreased volume of the relatively collapsed amniotic sac in oligohydramnios may twist or force the foot into the equino varus posture. Leg length discrepancy has also been noted in legs encircled by congenital constriction bands [27].

Extra-skeletal involvement may involve the face or abdomen. Though hideous cranial abnormalities are often incompatible with life, a number of children survive with severe facial abnormalities. The oblique facial clefts which occur as a result of amniotic bands are different from those which occur as a result of failure of structures to migrate or converge in the midline since the more usual clefts represent a failure of structures to merge at the midline [17]. Oblique facial bands may involve the lip, palate, nose and eye.

Many amputations occur through the diaphysis of tubular bones. These diaphyseal

amputations are different from the short hypoplastic digits which are the result of a failure of formation. Overgrowth of diaphyseal amputations is a frequent problem in acquired childhood amputations but never a problem in children with congenitally malformed hands. Overgrowth and symptomatic diaphyseal amputation ends are, however, occasionally seen in children with congenital constriction band syndrome. Thus, *in utero* diaphyseal amputations create some of the same problems observed in later, postnatal amputations in children. Burgess has documented brachydactyly of acrosyndactylized digit, suggesting an interference with normal phalangeal growth proximal to the site of tether [28].

SUMMARY

Congenital constriction band syndrome may present with a number of interrelated manifestations, including amputation, syndactyly and indentation. Review of a stillborn specimen and clinical observations suggests that these seemingly disparate abnormalities are all the logical result of intrauterine trauma to the embryologically defined hand. The resultant limbs demonstrate evidence of disruption of growth as well as deformity of portions of the residual limbs. Surgical treatment demands release of deep as well as superficial structures and may require multiple stages.

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REFERENCES

1. Patterson, T. J. S. Congenital Ring-Constriction. Br. J. Plast. Surg. 14:1–31, 1961.
2. Light, T. R. Les brides amniotiques - Physiopathologie et traitement. In: Les Malformations Congénitales du Membre Supérieur, Gilbert, A., Buck-Gramcko, D. and Lister, G., eds., Groupe d'Étude de la Main Monograph, Expansion Scientifique Française Paris, 18:85–91, 1991.
3. Buck-Gramcko, D. Congenital malformations. In: Hand Surgery. Buck-Gramcko, D., Millesi, H. and Lister, G. D., Eds. Georg Thieme Verlag, Stuttgart, 1988, VI, pp. 1242–1248.
4. DiMeo, L., Mercer, D. H. Single-stage Correction of Constriction Ring Syndrome. Ann. Plast. Surg. 19:469–474, 1987.
5. Hall, E. J., Johnson-Giebina, R. and Vasconez, L. O. Management of the Ring Constriction Syndrome: a reappraisal. Plast. Reconstr. Surg. 69:532–536, 1982.
6. Flatt, A. E. Constriction Ring Syndrome, Chapter 11 in The care of congenital hand anomalies. St. Louis, C. V. Mosby, 1977, pp. 213–227.
7. Upton, J. Correction of constriction rings. J. Hand. Surg. 16A:947–953, 1991.
8. Emmett, A. J. J. The ring constriction syndrome. Handchir. Mikrochir. Plast. Chir. 24:3–15, 1992.
9. Tada, K., Yonenobu, K., and Swanson, A. B. Congenital constriction band syndrome. J. Pediat. Orthop. 4:726–730, 1984.
10. Weeks, P. M. Radial, median and ulnar nerve dysfunction associated with a congenital constriction band of the arm. Plast. Reconstr. Surg. 69:333–337, 1982
11. Dobyns, J. H. Congenital Ring Syndrome (Congenital Constriction Band Syndrome) in Green D. P., ed. Operative Hand Surgery, 2nd edition. New York, Churchill Livingston, 1988, pp. 503–509.
12. Walsh, R. J. Acrosyndactyly: A study of twenty-seven patients. Clinical Orthopaedics and Related Research 71:99–111, 1970.
13. Moore, M. H. Nonadjacent syndactyly in congenital constriction band syndrome. J. Hand Surg. 17A:21–23, 1992.
14. Visuthikosol, V. and Hompuem, T. Constriction band syndrome. Annals of Plastic Surgery 21(5):489–495, 1988.
15. Miura, T. Congenital constriction band syndrome. J. Hand Surg. 9A:82–88, 1984.
16. Ogino, T. and Saitou, Y. Congenital constriction band syndrome and transverse deficiency. J. Hand Surg. 12B:343–348, 1987.
17. Van Helmont, J. B. Of material things injected or cast into the body. In: Ortus Medicinae, Amsterdam, Ludwig Elzevier, 1652, pp. 597–603.
18. Montgomery, W. F. Observations on the spontaneous amputation of the limbs of the fetus in

utero, with an attempt to explain the occasional cause of its production. Dublin Med. Chem. Sci. J. 1:140–144, 1832.
19. Streeter, G. L. Focal deficiencies in fetal tissues and their relation to intrauterine amputations. Carnegie Institute Contribs. Embryology 22:1–44, 1930.
20. Torpin, R. Amniochorionic mesoblastic fibrous strings and amnionic bands associated constricting fetal malformations or fetal deaths. Am. J. Ob. & Gyne. 91:65–75, 1965.
21. Torpin, R. and Faulkner, A. Intrauterine amputation with the missing member found in fetal membranes. JAMA 198:205–207, 1966.
22. Torpin, R. Fetal malformations caused by amnion rupture during gestation. Springfield, Charles C. Thomas, 1968.
23. Kino, Y. Clinical and experimental studies of the congenital constriction band syndrome with an emphasis on its etiology. JBJS 57A:636–643, 1975.
24. Higginbottom, M. C., Jones, K. L., Hall, B. D. and Smith, D. W. The amnionic band disruption complex: timing of amnionic rupture and variable spectra of consequent defects. J. Pediatrics 95:944–949, 1979.
25. Moses, J. M., Flatt, A. E. and Cooper, R. R. Annular constricting bands. JBJS 59A:562–565, 1977.
26. Cowell, H. R. and Hensinger, R. N. The relationship of club foot to congenital annular bands. In: Bateman, J.E. ed. Foot Science. Philadelphia, W. B. Saunders, 1976, pp. 41–46.
27. Askins, G. and Ger, E. Congenital constriction band syndrome. J. Pediatric. Orthop. 8:461–466, 1988.
28. Mayou, B. J. and Fenton, O. M. Oblique facial clefts caused by amniotic bands. Plast. Reconstr. Surg. 68:675–681, 1981.
29. Light, R. R. and Manske, P. Congenital hand malformations and deformities. In: Barr, J. S. Ed. AAOS Instructional Course Lecture Chicago, American Academy of Orthopaedic Surgeons, 38:31–71, 1989.