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### Neonatal Radial Nerve Palsy as a Manifestation of Congenital Ring Syndrome : A Case Report and Literature Review

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#### ABSTRACT

Constriction Ring Syndrome is a rare condition characterized by either partial or complete circular constrictions around limbs or digits. The manifestations of congenital band syndrome include syndactyly of digits, terminal amputations and digital edema distal to the constrictions. Occasionally, constriction may lead to injury of a peripheral nerve.

We report the case of a constriction ring syndrome in a newborn with radial nerve palsy caused by a band at the level of the arm. Acute release of congenital constriction band was performed releasing the band with neurolysis of the radial nerve. This led to decompression and the nerve restoration of hand function.

**Keywords:** Constriction band syndrome, Amniotic band syndrome, Radial nerve palsy

#### 1. Introduction

Constriction Ring Syndrome is a rare condition characterized by either partial or complete circular constrictions around limbs or digits. The manifestations of congenital band syndrome include syndactyly of digits, terminal amputations and digital edema distal to the constrictions. Occasionally, constriction may lead to injury of a peripheral nerve.

#### 2. Case Presentation

We report the case of a 1-day-old full term newborn, with a non-traumatic, spontaneous, vaginal delivery and with a birth weight of 3400 grams, who presented with congenital constriction band syndrome of the upper limb, causing radial nerve palsy. No maternal history of drug use or abuse reported

during pregnancy.

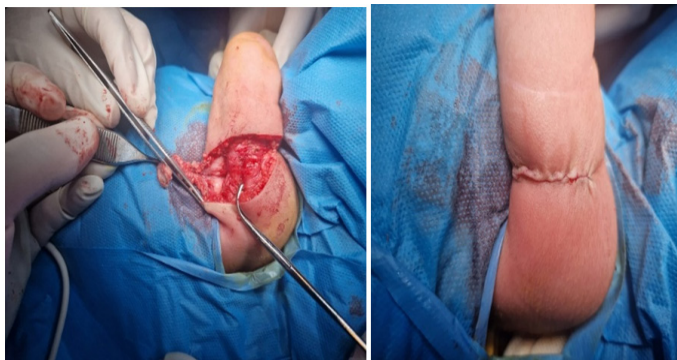
Physical examination revealed the presence of constriction band encircling the proximal third of the left upper arm with associated radial nerve palsy evident by clawing of the fingers. Hand perfusion was intact, with both radial and ulnar pulses easily palpable. Left shoulder and elbow function were fully preserved. The child had no additional abnormalities or comorbidities. There was no family history of similar anomalies. The tests for electromyography and nerve conduction were incomplete as the patient was unable to tolerate the examination.

A complete circumferential excision was performed, removing the constriction band and underlying fibrous tissue within the subcutaneous layer and fascia, along with a 1-2 mm

margin of surrounding healthy tissue. The wound is closed with standard side-to-side fashion. In our case, the early intervention was successful after decompression of the nerve with complete recovery of hand function without sequelae (Figures 1 and 2).



**Figure 1:** Constriction band in the proximal third of the left upper arm causing radial nerve palsy, characterized by clawing of the fingers.



**Figure 2:** Intraoperative photo demonstrating the complete circumferential excision of the constriction band.

### 3. Discussion

Congenital ring syndrome is a rare condition that may present with a variety of clinical manifestations, characterized by partial or complete circumferential constrictions around limbs or digits<sup>1</sup>. Consequences include acrosyndactyly of the digits, terminal amputations and localized swelling accompanied by digital edema distal to the constriction sites.

This syndrome is also known by various other names, including Amniotic Band Syndrome (ABS) or congenital constriction band, Amniotic band sequence, Amniotic Disruption Sequence, Streeter's dysplasia, pseudoainhum and annular grooves or defects, ADAM complex and intrauterine amputation. This condition is a common cause of terminal congenital limb malformations, with a reported incidence of 1 in 15,000 births and it is estimated to be responsible for 178 in 10,000 miscarriages<sup>2</sup>. It affects both sexes equally and there is no evidence of hereditary transmission.

### 4. Etiology

Despite the existence of multiple theories, the etiopathogenesis of constriction band syndrome remains unclear. There exist two principal conflicting theories concerning whether the initiation is intrinsic or extrinsic to the embryo or fetus.

The intrinsic theory, proposed by George Streeter, suggests that a defect in the subcutaneous germplasm leads to soft tissue necrosis followed by healing, resulting in the formation

of constriction bands<sup>3</sup>. According to this theory, a disruptive event occurs during blastogenesis, causing soft tissue to slough off without the involvement of amniotic bands. Subsequent external healing of the affected area leads to the development of constricting rings, which cause localized developmental defects.

The extrinsic theory, which is the most widely accepted, was proposed by Richard Torpin. He suggested that entanglement of the limbs in amniotic defects or free amniotic strands leads to constriction band syndrome, a condition from which many of the associated eponyms have originated. Following rupture, the amniotic sac ceases to grow properly and eventually separates from the chorion<sup>4</sup>. The chorionic side of the amnion gives rise to numerous mesoblastic fibrous strands that entangle and trap fetal parts. This entrapment causes constriction of the affected body part, disrupting blood flow to the area and potentially resulting in fetal amputations among other complications.

### 5. Clinical Features

The prenatal history may include oligohydramnios, premature uterine contractions and leakage of fluid. However, the mother often reports an uncomplicated pregnancy<sup>5</sup>. Although prenatal diagnosis by ultrasonography is occasionally possible, for most parents the affected child comes as a surprise. Newer techniques, such as three-dimensional ultrasonography, may improve prenatal diagnosis and high-resolution ultrasound, as well as improved fetoscopic surgical techniques, may eventually allow in utero surgical treatment of amniotic bands.

The clinical presentation of congenital constriction band syndrome varies widely among patients, depending on the severity and depth of the constriction<sup>6</sup>. These constrictions can range from superficial and incomplete bands to deep, circumferential rings that may nearly cause congenital amputations. Typically, the bands are located distally on the limbs; however, multiple constrictions can occur within the same limb. Involvement of the upper extremity is more common than that of the lower extremity. The digits are the most frequently affected part of the limbs, especially the longest central three.

Amputations commonly occur when constriction bands completely obstruct blood flow to a fetal limb, inhibiting its growth and resulting in tissue necrosis. Fenestrated syndactyly represents the next most frequent anomaly, occurring in nearly half of all cases. Constriction bands may lead to fusion of digits - either adjacent or nonadjacent - resulting in complex syndactyly, acrosyndactyly or both. This often presents as distal fusion of the digits accompanied by a proximal sinus between them.

Syndactyly, hypoplasia, brachydactyly, symphalangism, symbrachydactyly and camptodactyly have been reported in 80% of patients with congenital ring syndrome<sup>7</sup>.

Significant neurovascular impairment may be present distal to the constriction band. Impaired venous and lymphatic drainage causes swelling of the limb distal to the constriction. With growth, the constriction band occasionally gets more severe and becomes symptomatic. When constriction bands cross the body, they may result in a congenital fissure of the chest wall, a condition known as thoracoschisis. Nerve palsies have been associated with constriction ring sequelae and are present from birth<sup>9-10</sup>. These palsies, if explored surgically, are associated with absence of the nerve distally.

Around 7% of children with constriction band syndrome

present with craniofacial anomalies, including cleft lip and cleft palate. The prevalence of clubfoot in constriction band syndrome ranges from 12% to 56%. These deformities are frequently rigid and resistant to treatment, often involving a paralytic component due to peroneal muscle weakness and are always linked to ipsilateral constriction bands.

Angular deformities, bone dysplasia and pseudarthrosis may develop beneath constriction bands in both the upper and lower extremities. Additionally, leg-length discrepancy is observed in approximately 25% of patients.

## 6. Treatment

Because in most cases the amputation has occurred before delivery, it is rare that a newborn has an impending amputation that can be saved by the surgeon<sup>11</sup>. Belfort and colleagues have reported on fetoscopic release of an amniotic band on the lower leg of an infant in utero with exceptional results, but this technology and approach is still in its infancy. More often the newborn has a dried, necrotic part that cannot be saved<sup>12</sup>. This tissue may be surgically removed or allowed to slough. For incomplete congenital constriction bands without associated distal lymphedema, surgical intervention is generally not required, unless performed for cosmetic purposes.

There are different clinical scenarios where congenital constriction bands require surgical treatment: acutely in a neonate to salvage a limb or digit, deep bands that are causing vascular and/or neurologic compromise and cosmetic release and reconstruction for superficial bands.

In some cases, urgent surgical release of congenital constriction bands is necessary in neonates when the limb or digit is significantly compromised. This procedure is best carried out using a dorsal incision to release the band, rather than employing more extensive methods such as Z-plasty or circumferential excision<sup>13</sup>. More expedient surgical intervention is required if there is clear neurological impairment of the limb caused by compression beneath the constriction band. Since assessing this in neonates and young children is challenging, careful monitoring of hand function by both the family and the surgeon is essential to detect subtle asymmetrical changes.

Deep congenital constriction bands should be surgically removed down to healthy tissue and the resulting defect closed using multiple Z-plasty techniques. When the constriction band fully encircles the limb, a safer method involves staged excision-removing one half of the groove and closing it with Z-plasty, followed by a second procedure 2 to 3 months later. Following release, both lymphedema and cyanosis tend to improve progressively over time<sup>14</sup>. Simple excision of the bands with simple everting closure generally is inadequate because circumferential scar contracture may occur. Traditionally, the constriction band is released through a staged procedure, with an interval of 6 to 12 weeks between surgeries. This method aimed to facilitate better healing and limit necrosis at the skin edges, considering the reduced venous and lymphatic flow. Deep constriction bands are often excised and reconstructed safely at a single procedure. When the procedure is done, the ring of abnormal skin must be taken out completely. The circumferential line is then closed in standard side-to-side fashion if enough tissue exists or is converted into generously sized Z-plasties for a tension-free closure<sup>15</sup>. Although two-layer closure is preferable

in areas with enough of the subcutaneous layer present, on the digits this is usually not practical and here we use a single-layer closure.

A severe proximal neurologic defect and peripheral nerve palsy underlying a constriction ring on the forearm or arm may occur<sup>16,17</sup>. The effectiveness of electrodiagnostic assessment is not clearly established. Early exploration and appropriate treatment of the nerve lesion are reasonably considered after 6 months of age, successful outcomes following nerve decompression have been reported.

## 7. Conclusion

Patients with congenital constriction bands present with a wide range of clinical features, depending on the depth and extent of the constriction. These bands may range from superficial and incomplete to deep and circumferential, sometimes approaching congenital amputation. In some cases, peripheral nerve palsy can occur beneath the constriction band. The primary therapeutic goals are to restore function and improve aesthetics. Surgical excision of the constriction ring along with the underlying subcutaneous tissue is a fundamental aspect of treatment. Early intervention is crucial to achieving optimal outcomes.

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