Neonatal compartment syndrome is a rare condition. Early diagnosis and timely surgical intervention are paramount to optimize outcome. Time to fasciotomy is the most important prognostic factor. The purposes of this study were to describe a case presentation of neonatal compartment syndrome associated with a compound birth presentation and to perform a literature review. In this case, the neonate’s fingers were noted to be present on maternal cervical examination 24 hours before delivery. The patient then was noted to have a sentinel skin lesion. A diagnosis of neonatal compartment syndrome was suspected, and she underwent urgent fasciotomy. Literature review identified a total of 60 patients from 26 studies. Most patients were managed operatively. All patients presented with a sentinel skin lesion, emphasizing the importance of this clinical sign in diagnosis. Manometry is not routinely performed and no standards are available for acceptable pressure gradients.

Neonatal compartment syndrome (NCS) is a rare condition that can lead to long-term sequelae including Volkmann contracture, limb length discrepancies, and nerve lesions.\(^1\) Potential etiologies are typically classified as extrinsic or intrinsic: extrinsic causes include mechanical compression, which may result from oligohydramnios, amniotic band constriction, birth trauma, umbilical cord abnormalities, or malpresentation, whereas intrinsic causes include hypercoagulable states that may lead to intra-arterial or intravenous thromboses.\(^2\) Many cases are misdiagnosed, leading to fasciotomy delay and resultant poor prognosis.

In this report, we present the case of a neonate with compartment syndrome of the upper extremity associated with compound presentation (presentation of a fetal extremity with the presenting part of the fetus in the birth canal). The aim of this report is to increase awareness of this rare condition and its association with compound presentation, to improve diagnosis and treatment.

**Case Report**

A 3-kg female newborn was delivered by vacuum at 40 weeks 3 days’ gestation with no congenital anomalies present. The perinatal course was complicated by maternal chorioamnionitis and a compound presentation with the neonate’s fingers palpable on cervical examination both 24 and 4 hours before delivery. Apgar scores were 7 and 9 at 1 and 5 minutes, respectively. At 2.5 hours of life, there was notable swelling of the right forearm, nonblanching erythematous lesions of the volar and dorsal forearm, and a 1 x 1-cm volar wrist bulla (Fig. 1A); the neonatology team was consulted to evaluate the newborn. An x-ray was requested, which revealed no acute fracture (Fig. 1B). At 4.5 hours of life, on serial examination, there were additional volar forearm bullae as well as dusky discoloration of the digits (Fig. 1C), which prompted a hand surgery consultation. Doppler radial and ulnar pulses were intact.

On hand surgery consultation, the neonate was noted to have marked swelling of the right forearm, nonblanching erythematous lesions of the volar and dorsal forearm, and volar forearm bullae noted with dusky discoloration of the digits. Manometry was...
performed, which revealed a compartmental pressure of 22 mm Hg with a delta pressure of 32 mm Hg (the patient’s blood pressure was 74/54 mm Hg). The physical examination together with a prolonged labor and compound presentation raised concern for a clinical diagnosis of forearm compartment syndrome. Given the clinical concern, the patient was taken for emergent fasciotomy of the right forearm and hand. No further preoperative work-up was indicated.

An S-shaped incision was performed starting at the wrist flexion crease and proceeding proximally to the level of the forearm near the elbow under tourniquet control. The antebrachial fascia was incised longitudinally from the lacertus fibrosis to the wrist flexion crease, revealing bulging of superficial flexor muscle bellies. The deep flexor compartment was then exposed through the ulnar side of the forearm, beginning at the mid to distal forearm and following the interval between flexor carpi ulnaris and flexor digitorum superficialis, allowing release of flexor digitorum profundus and flexor pollicis longus fascia. Here, the deep compartment muscles appeared dusky in color but were fasciculating on contact with cautery. Then, the fascia overlying pronator quadratus was released. The carpal tunnel and Guyon canal were decompressed and both median and ulnar nerves appeared viable. The dorsal forearm and hand were then released using straight longitudinal incisions, and healthy muscle was noted on compartment release. The tourniquet was deflated and hemostasis was achieved. The dorsal wounds were closed primarily, and the volar forearm wound was left open. The skin overlying the carpal tunnel and thenar and hypothenar muscles was closed primarily. No skin was excised.

On postoperative day (POD) 1, dusky (Fig. 2A) and subsequently developed superficial necrosis on POD 4 (Fig 2B). The necrosis was allowed to demarcate, and the wound was managed with wet to moist dressings 3 times daily. The patient’s course was otherwise uneventful. On the day of discharge (POD 5), the patient was referred to outpatient hand therapy. Her parents were educated regarding a home exercise program for finger and wrist range of motion and use of an orthosis during nap times. On POD 14, the neonate again presented to the emergency department with her parents and was admitted overnight with superficial cellulitis of the dorsal hand in addition to a small dorsal hand abscess that eventually cultured methicillin-sensitive Staphylococcus aureus (Fig. 2C). She received cefazolin and underwent bedside debridement of the necrotic superficial flexor muscle during admission. Two months later, the wound had fully healed, and the patient had improving mild contracture of the volar wrist (Fig. 2D).

The patient’s family provided written informed consent to publish this case report and its accompanying images. We adhered to the CARE case report guidelines (https://www.care-statement.org/).

Discussion

Neonatal compartment syndrome is a rare diagnosis that requires early recognition and intervention. A literature review of published cases of NCS was performed using PubMed with the search terms “neonatal” and “compartment syndrome,” “Volkmann’s contracture,” or “ischemic contracture.” Studies published in languages other than English and those without full texts

![Figure 1. Preoperative images. A Sentinel skin lesions noted at 2.5 hours of life. B X-ray of upper arm with no acute abnormalities. C Progression of skin lesions at 4.5 hours of life.](image1)

![Figure 2. Postoperative images. A On postoperative day (POD) 1, duskiness of the muscles was noted. B By POD 4, there was development of superficial necrosis that was allowed to demarcate. C On POD 14, the patient again presented with superficial cellulitis of the dorsal hand and a small dorsal abscess positive for methicillin-sensitive Staphylococcus aureus. D Two months after surgery, the patient’s wound had fully healed with improving mild contracture of the volar wrist.](image2)
| Case Author | Year | Location | Key Diagnostic Findings | Time of Diagnosis | Intervention and Time | Suspected Etiology | Outcome |
|-------------|------|----------|-------------------------|-------------------|-----------------------|-------------------|---------|
| Tsur et al 17 | 1980 | L upper extremity | Sentinel skin lesions, paralysis, progressed to dry gangrene | Age 7 d | Fasciotomy and escharotomy, within 24 hours of diagnosis | Prolonged intrauterine pressure from an amniotic band | Volkmann contracture |
| Christiansen et al 18 | 1983 | R forearm, bilateral lower extremities | Sentinel skin lesions | Several days after birth | Fasciotomy, >24 hours after birth | Sepsis | Unknown |
| Caouette-Laberge et al 19 (5 cases) | 1992 | L (2) and R (3) upper extremities | Sentinel skin lesions (5), paralysis (2) | Unknown (5) | Surgical debridement (1), splinting (4) | Unilateral cord compression (1), oligohydramnios (1), u/k (3) | Scar contracture (1), Volkmann contracture (5), bone growth abnormality (5), nerve palsy (3) |
| Christiansen et al 20 (2 cases) | 1992 | R hand and forearm (2) | Sentinel skin lesions (2) | 2 h (1) and several hours (1) after birth | Fasciotomy at 3 hours of birth (1), conservative management (1) | Compression trauma (1), u/k (1) | Volkmann contracture (1), none (1) |
| Armstrong and Page 21 (6 cases) | 1997 | L (5) and R (1) upper extremity | Sentinel skin lesions (6) | Day of birth (1), time of delivery (5) | Fasciotomy (1), splinting (5) | Compressive thrombosis | Scar and Volkmann contracture (6), bone growth abnormality (2), amputation (1) |
| Tsujino et al 22 | 1997 | R forearm | Sentinel skin lesions, paralysis | Unknown | Conservative management | Intrauterine compression | Bone growth abnormality |
| Leautë-Lebrêze 23 et al | 1998 | R forearm and hand | Sentinel skin lesions | Unknown | Conservative management | Compression from dead fetus (co-twin) | Volkmann contracture, nerve palsy |
| Silfen et al 24 | 2000 | R upper extremity | Sentinel skin lesions that progressed to necrosis | 1 d after birth | Escharotomy, debridement, and fasciotomy on day 2 of life | Oligohydramnios | Volkmann contracture, bone growth abnormality |
| Ragland et al 25 (24 cases) | 2005 | L (11) and R (13) upper extremity | Sentinel skin lesions (24) | Within 3 h after birth (1), unknown (23) | Only 1 patient was treated at 3 hours of age. The other 23 were seen between 1 month - 13 years of age. A few cases were complicated by preterm birth, fetal distress during delivery, and coagulation abnormality. | Volkmann contracture |
| Dahlin et al (2 cases) 26 | 2009 | Unknown | Sentinel skin lesions and paralysis (2) | Unknown (2) | Fasciotomy on age 3 d | Shoulder dystocia | Unknown |
| Dandurand et al 27 | 2009 | L forearm and arm | Sentinel skin lesions that progressed to skin necrosis | Age 2 d | Fasciotomy, unclear timing | Abnormal arm position in utero | Unknown |
| Allen et al 28 | 2010 | R arm | Sentinel skin lesions with digital tip necrosis | Age 1 wk | Fasciotomy and debridement of muscle and skin | Abnormal arm position in utero | Unknown |
| Nanda et al 29 | 2010 | R forearm and hand | Sentinel skin lesions | At birth | Fasciotomy within 12 h of life | Umbilical cord compression | Autoamputation of thumb, bone growth abnormality |
| Rios et al 30 | 2011 | L forearm | Sentinel skin lesions that progressed to skin and muscle necrosis, paralysis | Age 4 d | Surgical debridement at age 14 d | Instrument delivery | Volkmann contracture |
| Isik et al 31 | 2012 | R hand and forearm | Sentinel skin lesions | At birth | Fasciotomy, unclear timing | Compound presentation, Amniotic band at birth, preterm twin, respiratory distress | Nerve palsy |
| Planeq et al 32 | 2013 | L forearm | Sentinel skin lesions with necrosis | Within first hours of life | Fasciotomy, unclear timing | Compound presentation | Weakness |
| Van der Kaay et al 33 | 2013 | R lower limb | Sentinel skin lesions | Unknown | Decompressing incisions | Severe birth trauma | Equinus of foot |
| Agrawal et al 34 | 2014 | R hand | Sentinel skin lesions | Age 5 d | Fasciotomy at age 112 h | Compressive thrombosis | Nerve palsy, bone growth abnormality |
| Pavlidis et al 35 | 2014 | L forearm and elbow fold | Sentinel skin lesions, paralysis | At birth | Unknown | Prothrombotic disorder | Unknown |
| Bekmez et al 36 | 2015 | L forearm and hand | Sentinel skin lesions, paralysis | After 24 h of life | Fasciotomy, unclear timing | Reperefusion injury after treating spontaneous axillary artery thrombosis resulting from coagulopathy | Scar contracture |
| Martinovski et al 37 | 2015 | L forearm and hand | Sentinel skin lesions | Within 9 h after birth | Fasciotomy, unclear timing | Compound presentation | Unknown |
| Mehta and Agarwal 38 | 2015 | R forearm and hand | Sentinel skin lesions | Time of birth | Fasciotomy at 6 h of life | Arterial thrombosis | Bone growth abnormality, autoamputation |
| Badawy et al 39 | 2016 | R upper extremity | Sentinel skin lesions, equivocal compartment pressures | Days after birth | Fasciotomy on d 7 of life | Disseminated intravascular coagulation | Scar contracture, Volkmann contracture |
| Martin and Trebarne 40 | 2016 | L forearm and hand | Sentinel skin lesions | Within hours of life | Fasciotomy at 6 h of life | Compressive thrombosis | Volkmann contracture |
| Tetreault et al 41 | 2018 | L forearm and hand | Sentinel skin lesions | Within 24 h of life | Fasciotomy within 24 h of life | Unknown | Volkmann contracture |
| Belli et al 42 | 2019 | L lower limb | Sentinel skin lesions | Within hours of life | Fasciotomy at 5 h of life | Compound presentation | None |

*For reports with multiple cases, information is reported using (n), which refers to the number of cases. The most highly suspected etiology for each case is listed. Outcomes are largely classified as scar contracture, Volkmann contracture, bone growth abnormality, nerve palsy, and amputation.*
available were excluded. A total of 60 patients were identified from 26 studies (Table 1). Fifty-five cases involved the upper extremities, whereas 3 affected the lower extremities. All patients presented with a sentinel skin lesion, which emphasizes the importance of this clinical sign in diagnosis. Diagnostic signs included sentinel skin lesions that may range from skin discoloration to necrosis.³

Compartment syndrome in adults is classically diagnosed when compartment pressures are 30 mm Hg or more greater than diastolic blood pressure; however, they are not routinely measured in the neonate because no standards are available for acceptable pressure gradients in newborns.¹ In most cases, the diagnosis was based on clinical findings without the measurement of compartment pressure.

Most patients were managed with fasciotomy. Early fasciotomy is crucial to optimize future limb function⁴; however, the vast predominance of subcutaneous fat, the compartment also may not experience bone growth abnormalities (32 patients). Other complications included nerve palsies (23 patients), scar contractures (9 patients), and amputation or auto-amputation (6 patients).

Long-term treatment should be aimed at improving functional deficits or loss and may include contracture release, tenolysis, neurolysis, skin grafting, nerve grafting, tendon transfer, and free tissue transfer, depending on the severity of disease.⁵-⁶ Prolonged ischemia may result in the development of Volkmann contracture.⁷

The most widely used classification system described by Tsuge⁸ divides clinical presentations of Volkmann contractures into mild, moderate, and severe types. The mild type is defined by localized disease affecting 2 or 3 fingers with little to no nerve involvement, the moderate type by degeneration of most or all of the flexor digitorum profundus and flexor pollicis longus with some nerve impairment, and the severe type by degeneration of the entire flexor compartment with severe sensory deficits.

Treatment of Volkmann contractures is focused on soft tissue, bone reconstruction, or both. In mild to moderate contracture the flexor pronator slide procedure⁹,¹⁰ is the mainstay of treatment. This technique involves release of the wrist flexor and pronator origin off the medial epicondyle followed by sequential release of the distal flexor origins from the ulna and intermuscular septum, working from proximal to distal and ulnar to radial.¹¹ Free-functioning muscle flaps using the gracilis and latissimus dorsi muscles have been reported for reconstruction of severe contracture of the finger and wrist where patients lack active finger flexion and available tendon transfers.¹²,¹³,¹⁴

Bone procedures, such as skeletal shortening and joint arthrodesis, are performed to match the length of fibrous muscle, although this is not ideal in developing children who have already sustained ischemic injury to the growth plates.⁷ If possible, additional surgical interventions should be delayed to minimize donor site morbidity in newborns.¹⁴ Postponing procedures until 1 year of age has been suggested to allow for the additional growth of muscles and vessels.¹⁵ In addition, neonates who undergo surgery and general anesthesia may be at risk for developing long-term abnormalities in organ maturation and deficits in neurocognition.¹⁵

Early diagnosis and timely surgical intervention are paramount to optimize outcomes of NCS.

Compartment pressures are not routinely measured because they may be inaccurate in this demographic.¹⁴,¹⁵ Owing to the predominance of subcutaneous fat, the compartment also may not feel tense as it does in adult patients, leading to a delay in diagnosis.¹ Clinical examination is critical because sentinel skin lesions are the most important diagnostic findings. If clinical suspicion is high, emergency fasciotomy should be performed. Given the association of external compression and NCS, a compound presentation at birth should raise suspicion for concomitant NCS in the setting of any abnormal skin changes.

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