



Contents lists available at ScienceDirect

Journal of Hand Surgery Global Online

journal homepage: www.JHSGO.org

Case Report

Neonatal Compartment Syndrome and Compound Presentation at Birth

Abra H. Shen^{*} Ruth Tevlin, MD,^{*} Matthew D. Kwan, MD,[†] Oscar H. Ho, MD,[†] Paige M. Fox, MD, PhD^{*}^{*} Division of Plastic and Reconstructive Surgery, Stanford University School of Medicine, Palo Alto, CA[†] Kaiser Permanente Santa Clara Medical Center, Santa Clara, CA

ARTICLE INFO

Article history:

Received for publication January 9, 2020

Accepted in revised form April 6, 2020

Available online May 8, 2020

Key words:

Fasciotomy

Manometry

Neonatal compartment syndrome

Sentinel skin lesion

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.

Published by Elsevier Inc. on behalf of The American Society for Surgery of the Hand.

This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

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 × 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

Declaration of interests: No benefits in any form have been received or will be received by the authors related directly or indirectly to the subject of this article.

Corresponding author: Paige M. Fox, MD, PhD, Department of Surgery, Division of Plastic and Reconstructive Surgery, Chase Hand and Upper Limb Center, Stanford University School of Medicine, 770 Welch Road, Suite 400, Palo Alto, CA 94304.

E-mail address: pfox@stanford.edu (P.M. Fox).

<https://doi.org/10.1016/j.jhsg.2020.04.001>

2589-5141/Published by Elsevier Inc. on behalf of The American Society for Surgery of the Hand.

This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

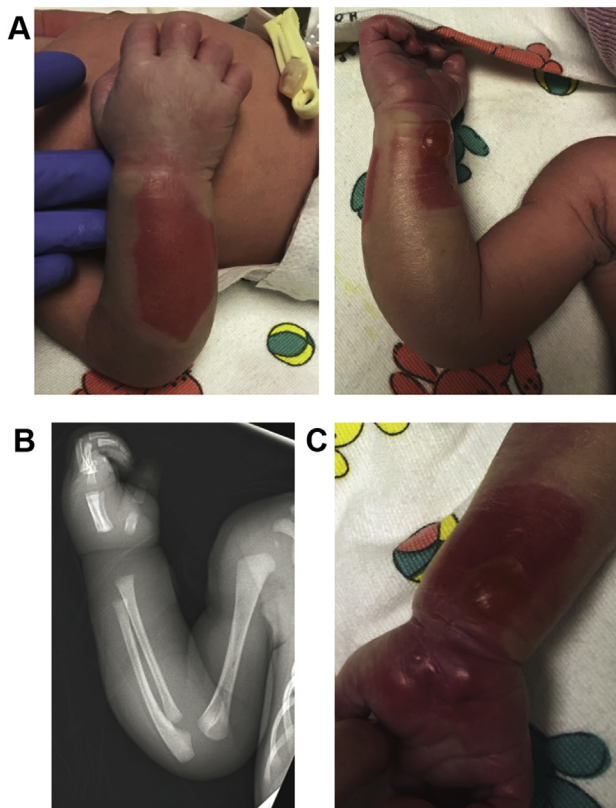


Figure 1. Preoperative images. **A** Sentinel skin lesions noted at 2.5 hours of life. **B** X-ray of upper arm with no acute abnormalities. **C** Progression of skin lesions at 4.5 hours of life.

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, the superficial flexor tendons were dusky (Fig. 2A) and subsequently developed superficial necrosis on POD 4 (Fig 2B). The necrosis was allowed to demarcate,



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.

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 resulting from the scar contracture with normal gliding of the flexor tendons (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," "Volkman's contracture," or "ischemic contracture." Studies published in languages other than English and those without full texts

Table 1
Summary of Literature Review*

Case	Author	Year	Location	Key Diagnostic Findings	Time of Diagnosis	Intervention and Time	Suspected Etiology	Outcome
1	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
2	Christiansen et al ¹⁸	1983	R forearm, bilateral lower extremities	Sentinel skin lesions	Several days after birth	Fasciotomies, >24 hours after birth	Sepsis	Unknown
3	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)	Umbilical cord compression (1), oligohydramnios (1), u/k (3)	Scar contracture (1), Volkmann contracture (5), bone growth abnormality (5), nerve palsy (3)
4	Kline and Moore ²⁰ (2 cases)	1992	L 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)
5	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)
6	Tsujino et al ²²	1997	R forearm	Sentinel skin lesions, paralysis	Unknown	Conservative management	Intrauterine compression	Bone growth abnormality
7	Léauté-Labrèze ²³ et al	1998	L forearm and hand	Sentinel skin lesions	Unknown	Conservative management	Compression from dead fetus (co-twin)	Volkmann contracture, nerve palsy
8	Silfen et al ²⁴	2000	R upper extremity	Sentinel skin lesions that progressed to necrosis	1 d after birth	Escharotomy, debridement, and then fasciotomy on day 2 of life	Oligohydramnios	Volkmann contracture, bone growth abnormality
9	Ragland et al (24 cases) ¹	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. Conservative management (2)	A few cases were complicated by preterm birth, fetal distress during delivery, and coagulation abnormality. u/k (2)	Volkmann contracture (15), bone growth abnormality (18), nerve palsy (16), amputation (3)
10	Dahlin et al (2 cases) ²⁵	2009	Unknown	Sentinel skin lesions and paralysis (2)	Unknown (2)	Conservative management (2)	u/k (2)	Bone growth abnormality (1), nerve palsy (1)
11	Dandurand et al ²⁶	2009	L forearm and arm	Sentinel skin lesions that progressed to skin necrosis	Age 2 d	Fasciotomy on age 3 d	Shoulder dystocia	Unknown
12	Allen et al ²⁷	2010	R arm	Sentinel skin lesions with digital tip necrosis	Age 1 wk	Fasciotomy and debridement of muscle and skin	Abnormal arm position <i>in utero</i>	Unknown
13	Nanda et al ²⁸	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
14	Rios et al ²	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
15	Isik et al ⁵	2012	R hand and forearm	Sentinel skin lesions	At birth	Fasciotomy, unclear timing	Compound presentation	Weakness
16	Plancq et al ²⁹	2013	L forearm	Sentinel skin lesions with necrosis	Within first hours of life	Fasciotomy, unclear timing	Amniotic band at birth, preterm twin, respiratory distress	Nerve palsy
17	Van der Kaay et al ³⁰	2013	R lower limb	Sentinel skin lesions	Unknown	Decompressing incisions	Severe birth trauma	Equinus of foot
18	Agrawal et al ³	2014	R hand	Sentinel skin lesions	Age 5 d	Fasciotomy at age 112 h	Compressive thrombosis	Nerve palsy, bone growth abnormality
19	Pavlidis et al ³¹	2014	L forearm and elbow fold	Sentinel skin lesions, paralysis	At birth	Unknown	Prothrombotic disorder	Unknown
20	Bekmez et al ³²	2015	L forearm and hand	Sentinel skin lesions, paralysis	After 24 h of life	Fasciotomy, unclear timing	Reperfusion injury after treating spontaneous axillary artery thrombosis resulting from coagulopathy	Scar contracture
21	Martinovski et al ³³	2015	L forearm and hand	Sentinel skin lesions	Within 9 h after birth	Fasciotomy, unclear timing	Compound presentation	Unknown
22	Mehta and Agarwal ³⁴	2015	R forearm and hand	Sentinel skin lesions	Time of birth	Fasciotomy at 6 h of life	Arterial thrombosis	Bone growth abnormality, autoamputation
23	Badawy et al ¹⁴	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
24	Martin and Treharne ⁴	2016	L forearm and hand	Sentinel skin lesions	Within hours of life	Fasciotomy at 6 h of life	Compressive thrombosis	Volkmann contracture
25	Tetreault et al ³⁵	2018	L forearm and hand	Sentinel skin lesions	Within 24 h of life	Fasciotomy within 24 h of life	Unknown	Bone growth abnormality
26	Belli et al ³⁶	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 pressures.

Most patients were managed with fasciotomy. Early fasciotomy is crucial to optimize future limb function⁴; however, the vast majority of patients did not undergo fasciotomy until after 24 hours of life. Other management routes included placement of an orthosis, decompressing incisions, and surgical debridement. Many patients experienced multiple complications and a total of 99 negative outcomes were reported, for an average of 1.65 negative outcomes reported per patient. One-third of patients experienced Volkmann contractures (33 patients) and almost one-third experienced 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.^{1,4–6} 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 procedure^{9,10} 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.^{1,12,13} Bone procedures, such as skeletal shortening and joint arthrodesis, are performed to match the length of fibrosed 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.¹² 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.^{14,16} 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.

References

- Ragland R, Moukoko D, Ezaki M, Carter PR, Mills J. Forearm compartment syndrome in the newborn: report of 24 cases. *J Hand Surg Am.* 2005;30(5):997–1003.
- Rios M, Ribeiro C, Soares P, et al. Volkmann ischemic contracture in a newborn. *BMJ Case Reps.* 2011;2011:bcr0520114201.
- Agrawal H, Dokania G, Wu SY. Neonatal volkmann ischemic contracture: case report and review of literature. *AJP Rep.* 2014;4(2):e77–e80.
- Martin B, Treharne L. Neonatal compartment syndrome. *Ann R Coll Surg Engl.* 2016;98(7):e111–e113.
- Isik C, Demirhan A, Karabekmez FE, Tekelioglu UY, Altunhan H, Ozlu T. Forearm compartment syndrome owing to being stuck in the birth canal: a case report. *J Pediatr Surg.* 2012;47(11):e37–e39.
- Poore SO, Bentz ML. The pediatric hand. In: Weinzweig J, ed. *Plastic Surgery Secrets.* Philadelphia: Mosby; 2010:776–782.
- Stevanovic M, Sharpe F. Management of established Volkmann's contracture of the forearm in children. *Hand Clin.* 2006;22(1):99–111.
- Tsuge K. Treatment of established Volkmann's contracture. *J Bone Joint Surg A.* 1975;57(5):925–929.
- Page CM. Four cases of flexion contracture of the forearm treated by a muscle-sliding operation. *Proc R Soc Med.* 1923;16(Sect Orthop):43–45.
- Scaglietto O. Sindromi cliniche immediate e tardive da lesioni vascolari nelle fratture degli arti [in Italian]. *Riforma Med.* 1957;71(27):749–755.
- Stevanovic MV, Sharpe F. Compartment syndrome and Volkmann ischemic contracture. 7th ed. In: Wolfe S, Pederson W, Kozen SH, Cohen M, eds. *Green's Operative Hand Surgery E-Book Vol 2.* Philadelphia, PA: Elsevier; 2016:1763–1787.
- Rubin G, Palti R, Gurevitz S, Yaffe B. Free myocutaneous flap transfer to treat congenital Volkmann's contracture of the forearm. *J Hand Surg Eur Vol.* 2015;40(6):614–619.
- Oishi SN, Ezaki M. Free gracilis transfer to restore finger flexion in Volkmann ischemic contracture. *Tech Hand Up Extrem Surg.* 2010;104–107.
- Badawy SM, Gust MJ, Liem RI, Ball MK, Gosain AK, Sharathkumar AA. Neonatal compartment syndrome associated with disseminated intravascular coagulation. *Ann Plast Surg.* 2016;76(2):256–258.
- Weiss M, Hansen TG, Engelhardt T. Ensuring safe anaesthesia for neonates, infants and young children: what really matters. *Arch Dis Child.* 2016;101(7):650–652.
- Hughes TB, Fowler J. Compartment syndrome and Volkmann ischemic contracture. In: Trumble TE, Rayan GM, Budoff JE, Baratz ME, Slutsky DJ, eds. *Principles of Hand Surgery and Therapy.* 3rd ed. Philadelphia, PA: Elsevier; 2016:180–192.
- Tsur H, Yaffe B, Engel Y. Impending Volkmann's contracture in a newborn. *Ann Plast Surg.* 1980;5(4):317–320.
- Christiansen SD, Desai NS, Pulito AR, Slack MR. Ischemic extremities due to compartment syndromes in a septic neonate. *J Pediatr Surg.* 1983;18(5):641–643.
- Caouette-Laberge L, Bortoluzzi P, Egerszegi EP, Marton D. Neonatal Volkmann's ischemic contracture of the forearm: a report of five cases. *Plast Reconstr Surg.* 1992;90(4):621–628.
- Kline SC, Moore JR. Neonatal compartment syndrome. *J Hand Surg Am.* 1992;17(2):256–259.
- Armstrong AP, Page RE. Intrauterine vascular deficiency of the upper limb. *J Hand Surg Eur.* 1997;22(5):607–611.
- Tsujino A, Hooper G. Neonatal compression ischaemia of the forearm. *J Hand Surg Eur.* 1997;22(5):612–614.
- Léauté-Labréze C, Depaire-Duclos F, Sarlangue J, et al. Congenital cutaneous defects as complications in surviving co-twins. *Arch Dermatol.* 1998;134(9):1121–1124.
- Silfen R, Amir A, Sirota L, Hauben DJ. Congenital Volkmann-Lesser ischemic contracture of the upper limb. *Ann Plast Surg.* 2000;45(3):313–317.
- Dahlin LB, Erichs K, Rosberg HE. Compartment syndrome in the forearms of two neonates. *Scand J Plast Reconstr Surg Hand Surg.* 2009;43(1):58–60.
- Dandurand M, Michel B, Fabre C, Stoebner P, Meunier L. Neonatal Volkmann's syndrome. *Ann Dermatol Venereol.* 2009;136(11):785–789.
- Allen LM, Benacci JC, Trane RN, Driscoll RJ. A case of neonatal compartment syndrome: importance of early diagnosis in a rare and debilitating condition. *Am J Perinatol.* 2010;27(2):103–106.
- Nanda R, Kanapathipillai P, Stothard J. Selective growth disturbance of the hand following neonatal compartment syndrome: a case report. *J Hand Surg Eur.* 2009;34(6):813–814.

29. Plancq MC, Buisson P, Deroussen F, Krim G, Collet LM, Gouron R. Successful early surgical treatment in neonatal compartment syndrome: case report. *J Hand Surg Am.* 2013;38(6):1185–1188.
30. Van Der Kaay DCM, Horsch S, Duvekot JJ. Severe neonatal complication of transverse lie after preterm premature rupture of membranes. *BMJ Case Rep.* 2013;2012–2014.
31. Pavlidis E, Spagnoli C, Duca M, Ormitti F, Magnani C, Pisani F. Neonatal forearm compartment syndrome: look for cerebral stroke. *J Pediatr.* 2014;164(2):427.e1.
32. Bekmez S, Beken S, Mermerkaya MU, Ozkan M, Okumus N. Acute forearm compartment syndrome in a newborn caused by reperfusion after spontaneous axillary artery thrombosis. *J Pediatr Orthop Part B.* 2015;24(6):552–555.
33. Martinovski M, Wilseck ZM, Mattson SE. Neonatal compartment syndrome due to compound birth presentation. *J Pediatr Surg Case Reports.* 2015;3(2):79–81.
34. Mehta S, Agarwal J. Neonatal compartment syndrome. *Eplasty.* 2015;15:ic57.
35. Tetreault AK, Axibal DP, Scott FA. Neonatal compartment syndrome treated within the first 24 hours of life. *Orthopedics.* 2018;41(5):e731–e733.
36. Belli G, Cucca G, Filippi L. Decompressive fasciotomy in an extremely preterm newborn with compartment syndrome. *J Pediatr.* 2019;214:232–233.e1.