Successful Early Surgical Treatment in Neonatal Compartment Syndrome: Case Report

M. C. Plancq, MD, P. Buisson, MD, F. Deroussen, MD, G. Krim, MD, L. M. Collet, MD, R. Gouron, MD, PhD

Neonatal compartment syndrome is rare, and the diagnosis is often missed or delayed because other ischemic diseases can mimic clinical signs observed on the skin. A premature newborn infant presented with skin lesions during the first hours of life that were recognized as the sentinel finding in compartment syndrome of the newborn. We restored normal function by emergency surgery. The authors highlight the importance of effective collaboration between pediatricians and surgeons to improve the management of this neonatal condition. *(J Hand Surg 2013;38A:1185–1188. Copyright © 2013 by the American Society for Surgery of the Hand. All rights reserved.)*

Key words Neonatal compartment syndrome, fasciotomy.

NEONATAL COMPARTMENT SYNDROME is rare, with only a few cases reported in the literature.[1–5] It is often associated with poor outcome because of delayed diagnosis. We report a case in which the diagnosis was established rapidly and emergency surgery was performed. This condition is difficult to diagnose during the neonatal period because other skin diseases such as gangrene of the newborn, necrotizing fasciitis, and aplasia cutis congenita can mimic compartment syndrome. Here, we discuss the clinical signs that may lead to diagnosis, and the limitations of medical imaging. The time from birth to surgery is the main prognostic factor.[6] Emergency surgery within hours of birth achieved good results.

CASE REPORT

A male infant, weighing 1,590 g, was born at 32 weeks of gestation after a twin bi-amniotic pregnancy induced by clomiphene. Pregnancy was complicated by premature labor treated by betamethasone. Premature rupture of membranes occurred at 30 weeks, requiring treatment with amoxicillin and clavulanic acid. These complications were also exacerbated by maternal nicotine addiction. Fetal respiratory distress at delivery required resuscitation and mechanical ventilation for 6 days. Several minutes after birth, the left forearm was erythematous with blisters, and we observed a constriction band on the proximal third of the forearm. Two hours later, the arm was cyanotic with blisters on the forearm and ischemia in the hand, but the fingers remained vascularized (Fig. 1). The lesions were extensive (blisters, then hemorrhagic blisters and necrotic skin) on the dorsal surface of the forearm, wrist, and hand. Doppler ultrasonography showed persistence of radial and ulnar vascular flows but suggested decreased blood flow compared with the left side. Emergency surgical decompression (skin incision and forearm compartments fasciotomy) revealed muscle ischemia despite incision of the constrictive band. We performed fasciotomy

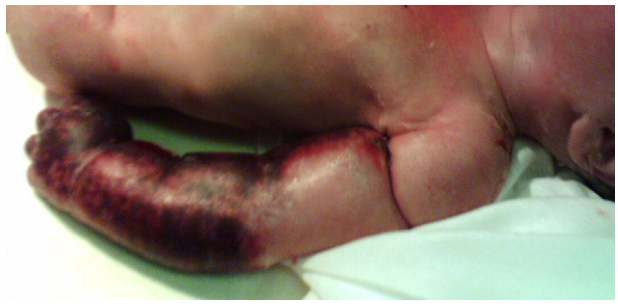

FIGURE 1: Appearance of the forearm 2 hours after birth.

From the Department of Pediatric Orthopedic Surgery and the Neonatal Intensive Care Unit, Amiens University Hospital, University of Picardie Jules Verne, Amiens, France.

Received for publication December 18, 2012; accepted in revised form March 8, 2013.

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

Corresponding author: Richard Gouron, MD, PhD, Department of Pediatric Orthopedic Surgery, Amiens University Hospital, Hôpital Nord Place Victor Pauchet, Amiens Cedex 1 80054, France; e-mail: richard.gouron@gmail.com.

0363-5023/13/38A06-0020$36.00/0
http://dx.doi.org/10.1016/j.jhsa.2013.03.029

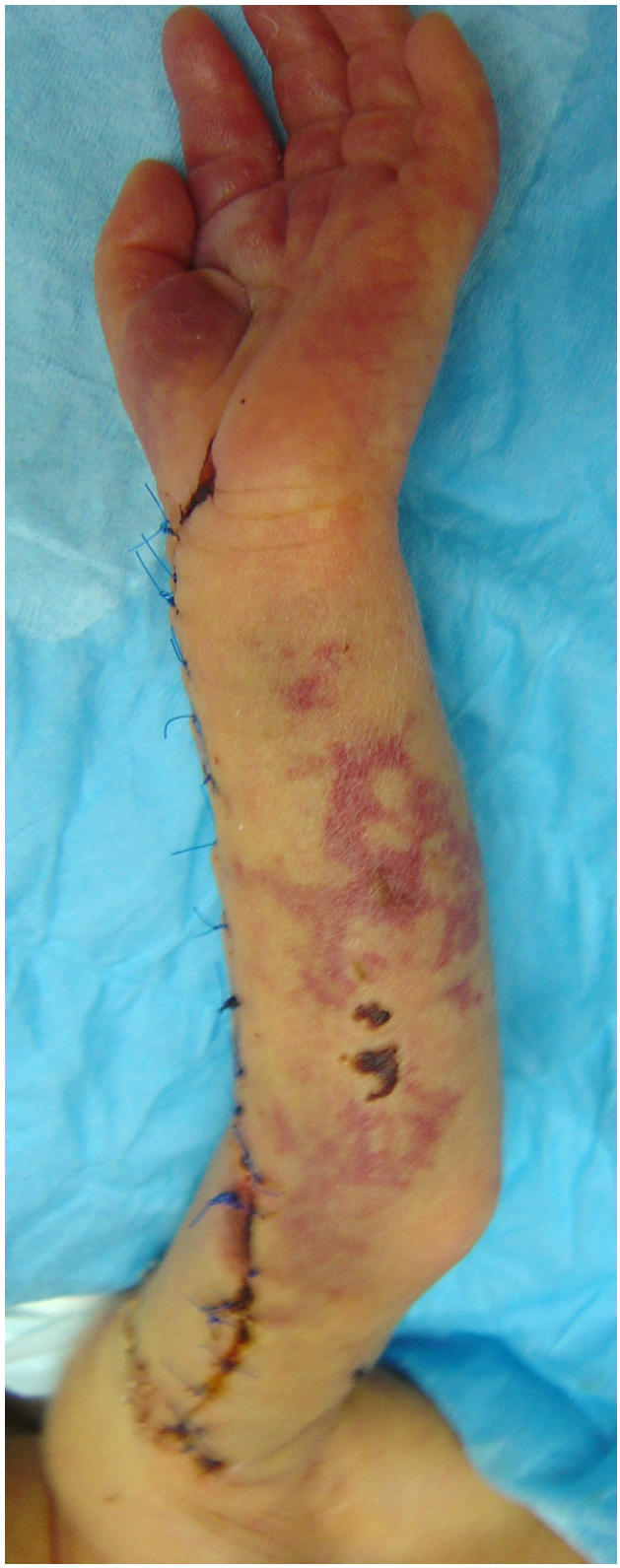

FIGURE 2: Appearance of the forearm after early fasciotomy, showing good muscle color.

from the upper arm to the wrist, allowing recovery of good muscle color (Fig. 2). After 4 days, the forearm and hand edema had decreased and the hand was well vascularized. We performed resection of the constriction band with double z-plasty. The skin edges of the fasciotomy were progressively closed, with good healing of the superficial necrotic areas (Fig. 3). The parents observed a certain degree of lack of sensation during the first 3 months of life, after which the child recovered complete forearm and hand function with no signs of contracture. Five years later, the limb presented normal function, with no clinical (Fig. 4) or radiographic (Fig. 5) growth anomalies compared with the right side.

DISCUSSION

The diagnosis of neonatal compartment syndrome in this case was based on the sudden onset of ischemia; the presence of skin blisters; the site of the lesion, typically involving the dorsal surface of the forearm, wrist, and hand; the absence of distal necrosis; and the good outcome in response to fasciotomy. The neonatal form of compartment syndrome is rare. Only a few cases have been reported in the literature.[4,7] In 2005, Ragland et al[6] published a series of 24 cases, often with poor results because of delayed diagnosis. Sequelae were Volkmann syndrome (71%), bone changes (75%), and nerve lesions (67%). This condition can be misdiag-

FIGURE 3: Good wound healing after z-plasty of the amniotic band constriction and closure of the skin defect 10 days after birth.

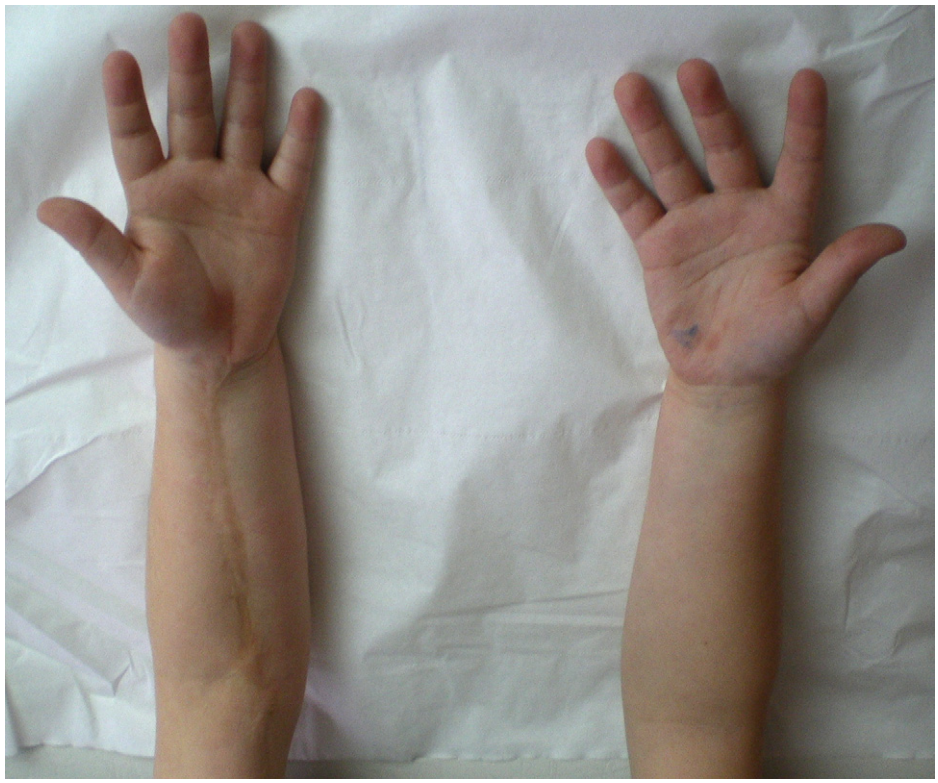

FIGURE 4: Appearance of the forearm 5 years later, compared with the right side, showing equal growth.

nosed because of its rarity and because skin lesions can mimic other ischemic diseases of the newborn. In gangrene of the newborn, lesions usually involve the distal extremities and often the lower limb.[8] This condition must be distinguished from necrotizing fasciitis, but the general condition and the fulminating course of sepsis and skin lesions usually guide the diagnosis. Aplasia cutis congenita, congenital absence of skin, is characterized by skin ulcers sometimes covered by a thin epithelial membrane at birth that usually heal spontaneously during the first weeks of life. These lesions often involve the scalp, trunk, and extremities, with a symmetrical distribution.[9]

Differential diagnosis is difficult because these diseases may stem from the same causes. Several compression factors and neonatal conditions can induce neonatal compartment syndrome. Local mechanical causes may be present: umbilical cord loops, fetal posture and oligoamnios, twin pregnancy, maternal uterine malformation, and amniotic band constriction.[4,6,7] This mechanical compression can be accentuated by maternal and neonatal conditions such as respiratory distress, vascular insufficiency, clotting disorders, and maternal diabetes.[4] Several of these factors were associated in the case reported here. The presence of an amniotic constriction band at birth was exacerbated by the neonatal condition (preterm twin, respiratory distress, and low cardiac output).

Compartment syndrome requires emergency surgical management, and therefore requires early recognition and cooperation between medical and surgical teams. Diagnosis and assessment of skin lesions can be difficult in small babies because they are sometimes sedated. The indication for surgery is guided by calculating the difference between diastolic blood pressure and compartment pressure, as recommended in adults. For many surgeons, fasciotomy is recommended when the pressure difference is less than 30 mm Hg.[10] However, in neonates, the mean diastolic blood pressure is about 40 mm Hg or less, a small increase in compartment pressure may thus be difficult to identify, and muscle viability may be already compromised.[6] Doppler ultrasonography can sometimes provide information about radial and ulnar blood flow that can contribute to the diagnosis.[3] However, these investigations must not delay surgical decompression. The procedure usually required is decompressive fasciotomy, and a good result can be expected when this is performed within the first hours after delivery, as in the case reported here.[1,4,6]

The prognosis of compartment syndrome is closely correlated with the time to diagnosis and early referral to the surgeon.[6] Effective collaboration between pediatricians and surgeons is essential to ensure assessment and early treatment. Most published cases were associated with late referral to the surgeon at the stage of irreversible

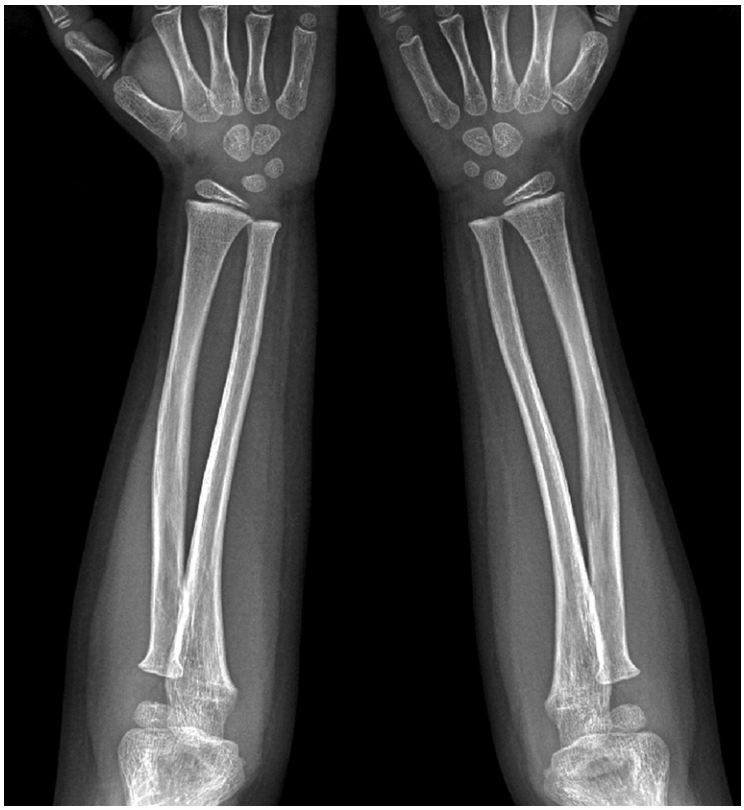

FIGURE 5: X-ray of the involved left forearm 5 years later, compared with the uninvolved side, showing no bone change and no growth arrest.

muscle damage responsible for Volkmann syndrome, with contractures and major functional sequelae including nerve dysfunction and growth disorders of the forearm.[4–6] The long-term results are disappointing, with muscle contracture and growth disorders of the radius and ulna that can cause deformities.[2,5,11] Rehabilitation and secondary surgery can limit the consequences of Volkmann syndrome, but the functional results are poor.[9] Long-term follow-up is required. Early recognition and treatment may obviate these problems.

REFERENCES

1. Allen LM, Benacci JC, Trane RN III, 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.
2. Dahlin LB, Erichs K, Rosberg H-E. Compartment syndrome in the forearms of two neonates. *Scand J Plast Reconstr Surg Hand Surg.* 2009;43(1):58–60.
3. Cham PMH, Drolet BA, Segura AD, Esterly NB. Congenital Volkmann ischaemic contracture: a case report and review. *Br J Dermatol.* 2004;150(2):357–363.
4. Goubier JN, Romaña C, Molina V. [Neonatal Volkmann's compartment syndrome: a report of two cases]. *Chir Main.* 2005;24(1):45–47.
5. Nanda R, Kanapathipillai P, Stothard J. Selective growth disturbance of the hand following neonatal compartment syndrome: a case report. *J Hand Surg Eur Vol.* 2009;34(6):813–814.
6. Ragland R III, 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.
7. Dandurand M, Michel B, Fabre C, Stoebner P, Meunier L. [Neonatal Volkmann's syndrome]. *Ann Dermatol Venereol.* 2009;136(11):785–789.
8. Hensinger RN. Gangrene of the newborn: a case report. *J Bone Joint Surg Am.* 1975;57(1):121–123.
9. Léauté-Labrèze C, Depaire-Duclos F, Sarlangue J, et al. Congenital cutaneous defects as complications in surviving co-twins: aplasia cutis congenita and neonatal volkmann ischemic contracture of the forearm. *Arch Dermatol.* 1998;134(9):1121–1124.
10. Mars M, Hadley GP. Raised compartmental pressure in children: a basis for management. *Injury.* 1998;29(3):183–185.
11. Tsujino A, Hooper G. Neonatal compression ischaemia of the forearm. *J Hand Surg Br.* 1997;22(5):612–614.