Congenital and Perinatal Compartment Syndrome: The Things We Have Learned

Ann. Ital. Chir., 2026 97, 11: 1942–1945
https://doi.org/10.62713/aic.4869

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To the Editor,

Congenital and perinatal compartment syndrome of the upper extremity represents a rare but clinically relevant condition where uncertainty exists about treatment regimens. Sometimes it could even be misdiagnosed as aplasia cutis congenita (aplasia cutis congenita [ACC] type VII), potentially leading to a delay in appropriate surgical treatment. Given the overlapping clinical presentation and the limited number of reported cases, the distinction between congenital compartment syndrome and ACC type VII remains challenging and often depends on the clinical experience of the initial examiner. We initially had difficulty distinguishing congenital compartment syndrome from ACC type VII in one patient, and would also like to contribute to this discussion by critically re-evaluating the concept of proposed ACC type VIIb. Aplasia cutis congenita is a heterogeneous disorder characterized by localized absence of skin, classically classified according to Frieden [1] into nine subtypes, with type VII involving the skin of the extremities. A later subdivision into type VIIa and VIIb, with associated bony or neurological abnormalities, has been proposed [2,3]. From a surgical perspective, these additional features are highly suggestive of an ischemic incident rather than a primary developmental skin defect, thereby raising the question whether ACC type VIIb constitutes a distinct entity at all.

In our institutional experience over 13 years, we identified three cases of congenital and perinatal compartment syndrome, one of which was initially misdiagnosed as ACC type VII. The cases were identified through the institutional medical record system, in which patients with congenital hand malformations were registered separately. No formal database search or diagnostic-code-based search was performed. We define perinatal compartment syndrome as an acute clinical presentation characterized by signs of edema, pain, swelling, tenderness, and a flexion contracture of the wrist and/or fingers that is still correctable, occurring during or after delivery. In contrast, congenital compartment syndrome already shows signs of defect healing. Here, we would like to share our experiences.

A narrative literature search was conducted using PubMed, Web of Science, Scopus, and Google, supplemented by screening the reference lists of relevant publications. The search terms included “aplasia cutis congenita”, “congenital compartment syndrome”, “neonatal compartment syndrome”, “congenital Volkmann contracture”, “pediatric”, “child”, and “neonatal”. No restrictions were imposed on the publication date. Publications directly relevant to the topic were included. TW screened the retrieved literature, extracted the relevant information, and performed the literature analysis. This was a narrative review rather than a systematic review.

The first case involved an extremely premature neonate (female; 650 g; gestational age 23 weeks and 6 days; otherwise healthy; umbilical cord prolapse) who developed compartment syndrome following arterial line extravasation on day 7 after birth. Subsequently, she developed defects of the skin and deeper structures of her right forearm. Our department was consulted three weeks after birth because of stagnating wound healing and discoloration from wrist to elbow after massive swelling. We diagnosed her with a missed perinatal compartment syndrome. During our first plastic surgery assessment, we found a flexed wrist and clawing position of the fingers, with minimal passive extension and a reduced diameter of the affected forearm compared to the healthy side. Palpation of the forearm caused pain during finger extension. Considering the patient’s age and physical size, as well as the signs of recovery, we have decided to continue with non-surgical treatment. After conservative treatment with hydrocolloid dressings, which were changed once weekly, and intensive hand therapy with stretching, the wounds healed completely in two weeks. The claw position was much improved with complete active extension of the fingers and a full wrist range of motion. One week after complete wound healing, we found a thumb still in an adducted position at rest, but capable of active abduction. Three weeks after this assessment, this improved to

Submitted: 25 June 2026 Revised: 15 August 2026 Accepted: 21 August 2026 Published: 10 November 2026
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Editor: Roberto Cirocchi

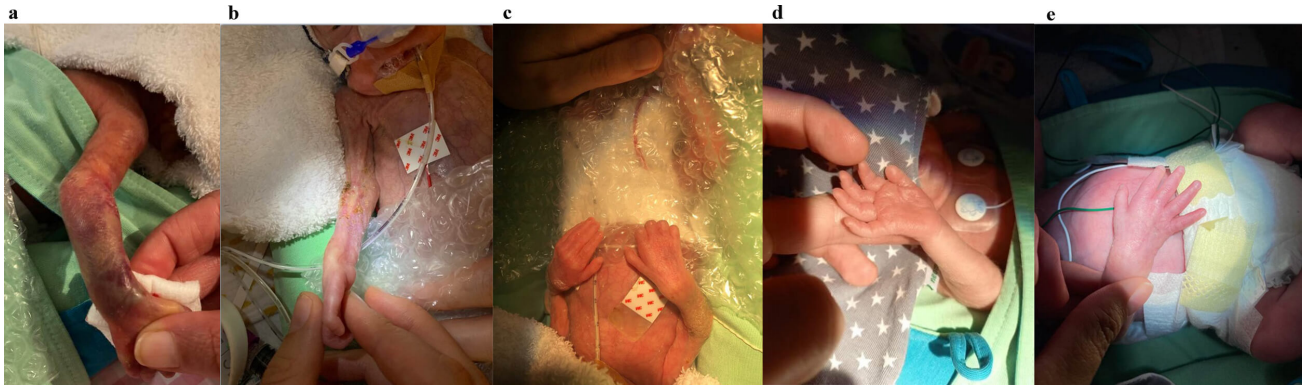


Fig. 1. Neonate with an early congenital compartment syndrome. One week after birth with skin necrosis due to an evident compartment syndrome after extravasation of an arterial line in the right arm (a). Two weeks after birth: healing skin and a beginning claw position of the fingers (b). Three weeks after birth: a claw position of the right hand compared to the uninvolved left hand/forearm (c). Seven weeks after birth: a complete reconstitution of the skin and function under conservative treatment (d,e).

only slightly reduced thumb abduction. Despite delayed diagnosis, complete functional and morphological recovery was observed over a two-year follow-up period with conservative management consisting of occlusive wound therapy and intensive physiotherapy (Fig. 1), without surgery.

The second case concerned a premature neonate (male gemelli; 2333 g; pH 6.86; base excess –17.5 mmol/L; gestational age 36 weeks and 2 days; borne via vacuum extraction under maternal pre-eclampsia with a 2 × 2 cm large white/yellow blister on his right forearm; minimal hypospadias; otherwise healthy) initially diagnosed with ACC type VII based on a sharply demarcated skin lesion at the forearm. Delayed subsequent ultrasound imaging -on advice of the plastic surgery department- demonstrated fibrosis of the deep flexor compartment, consistent with prenatal ischemia. The ultrasound showed a hyperechoic appearance of the deep flexor compartment in the right forearm, atrophy of the subcutaneous tissue and pronator teres muscle just distal to the right elbow, hyperechogenicity of the distal superficial flexors on the right, and atrophy of the deep thenar compartment on the right. The deep extensors of the fingers and thumb also showed abnormal appearances, whereas the brachioradialis retained relatively normal muscle bulk and appearance. The diagnosis was therefore revised to congenital compartment syndrome. With these findings, the delayed surgical consultation, and the positive clinical course, no further testing or imaging was initiated. During conservative management and intensive hand physiotherapy, the patient showed progressive functional recovery, with only minor residual impairment of thumb abduction at the 2-year follow-up (Fig. 2).

The third case highlights the importance of early surgical intervention. A premature neonate (male; 2620 g; gestational age 34 weeks and 5 days; otherwise healthy) developed acute compartment syndrome secondary to spontaneous hemorrhage in the context of enterovirus-induced liver failure. He had spontaneous hemorrhage with consecutive compartment syndrome of his left forearm one week after birth. Clinical signs of a compartment syndrome were an extensive hematoma and painful swelling from the elbow to the hand with light flexion of the wrist and fingers. We performed surgery 10 days after birth, on the day of the first plastic surgery consultation- a carpal tunnel and Guyon's canal release, and forearm volar and dorsal fasciotomies through multiple small incisions immediately after optimization of his coagulation. Intraoperatively, the muscles were all vital. Complete functional recovery was observed within two months after surgical decompression, despite initially delayed postoperative progress due to coagulopathy. Functional recovery was assessed clinically through observation of normal joint range of motion and functional movement patterns, without the use of standardized functional assessment tools or objective quantitative measurements. The patient was transferred to another hospital for liver transplantation and lost to further follow-up (Fig. 3).

These cases illustrate several important aspects. Most importantly, the timing of onset appears to be a decisive prognostic factor. Early intrauterine onset may allow for partial or even complete regeneration due to embryonic tissue plasticity, whereas late prenatal or perinatal onset requires timely therapeutic intervention to prevent irreversible damage. Based on our three case studies, we draw the following conclusions: First, in cases of acute clinical presentation with progressive symptoms involving wrist/finger flexion contracture, pain, tenderness, and swelling, urgent surgical evaluation and timely compartment release should be considered, unless intervention must be delayed to address other life-threatening conditions (e.g., a coagulopathy). Second, in cases of delayed consultations, where signs of healing are already present and acute symptoms have subsided, conservative treatment can also lead to favorable results in newborns depending on the gestational age. Although clinical functional recovery was observed during



Fig. 2. Neonate with a late congenital compartment syndrome. On the day of birth, ith a skin defect and flexion contracture of the right wrist (a). Six weeks later, following conservative treatment and hand therapy (b). Five months later, with progressive functional recovery (c). At 16 months (d) and 26 months after birth (e).



Fig. 3. Neonate with a perinatal compartment syndrome. Initial findings at the first plastic surgery consultation (dorsal and palmar aspect of the left forearm) ten days after birth (a,b). One week after compartment release, swelling was reduced, but the hematoma was still extensive (c,d). Two months after compartment release of his left hand/forearm, a normal finger cascade is evident (e,f).

follow-up, a reliable standardized functional assessment in children is only possible starting at around four years of age; therefore, formal functional assessment results were unavailable for our patients. Third, the presence of associated bony abnormalities, neurological deficits, or muscle fibrosis as described in proposed ACC type VIIb is more consistent with ischemic pathophysiology and growth disturbance, as observed in compartment syndrome [4]. Therefore, we believe that ACC type VIIb is most probably a misnomer and, in our opinion, is unlikely to exist; this assessment is based solely on our own clinical experience and should also be validated through larger case series and further investigations. Fourth, measuring compartment pressure offers no additional clinical benefit. If a persistent acute process is suspected, surgical decompression is still necessary.

A review of the literature reveals that only two cases have been reported as ACC type VIIb [2,3], and these descriptions likely represent misclassified congenital compartment syndromes. Additional reports classified as ACC similarly meet criteria for ischemic injury [5]. In agreement with Léauté-Labrèze et al. [6], true ACC type VII should not involve deeper structural abnormalities or functional impairment. Also, bilateral involvement has been reported in cases of ACC type VII, except in one puplished case. Conversely, congenital Volkmann ischemic contracture and neonatal compartment syndrome consistently present with unilateral involvement, cutaneous necrosis or ulceration, and variable degrees of motor impairment [7,8,9,10,11,12,13,14,15].

Based on these observations, we propose that ACC type VIIb should not be maintained as a separate diagnostic category. Instead, such cases should be considered within the spectrum of congenital and perinatal compartment syndrome. Accurate and early diagnosis is essential, as both conservative and surgical strategies can lead to excellent outcomes when applied appropriately, whereas delayed recognition may result in permanent functional deficits.

Availability of Data and Materials

The data that support the findings of this study are available from the corresponding author upon reasonable request.

Author Contributions

TW and ASK designed this study. TW conducted the literature screening, data extraction and data analysis. TW wrote the manuscript. ASK, SH and DU participated in the discussion of this report, interpreted key data from the study, and contributed to data verification and proofreading. All authors contributed to the critical revision of the manuscript

for important intellectual content. All authors read and approved the final manuscript. All authors have participated sufficiently in the work and agreed to be accountable for all aspects of the work.

Ethics Approval and Consent to Participate

This study involved only a retrospective analysis of routine clinical data, and no additional interventions were carried out. For this type of study, formal ethical approval from the local ethics committee (CMO Radboudumc) is not required. The parents of the children gave informed consent. This study complies with the Declaration of Helsinki.

Acknowledgment

Not applicable.

Funding

This research received no external funding.

Conflict of Interest

The authors declare no conflict of interest.

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