Congenital Volkmann Syndrome and Aplasia Cutis of the Forearm
A Challenging Differential Diagnosis

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Differential diagnosis between congenital Volkmann ischemic contracture (CVIC) and unilateral aplasia cutis congenita (ACC) type VII of the forearm presents a clinical challenge. Both diseases share the same clinical presentation characterized by a stellate ulceration over the upper extremities and reported association with neuromuscular defects, but the diagnostic criteria to differentiate these 2 entities remain unclear.

OBSERVATIONS A newborn girl presented with an ulceration of the left forearm associated with an apparent neurological impairment. On the basis of the suspected neurological involvement, a diagnosis of CVIC was initially considered, but because the neurological evaluation excluded paralysis, our final diagnosis was ACC type VII.

CONCLUSIONS AND RELEVANCE In our opinion, CVIC should be considered a particular form of ACC in which an external noxa affects the forearm, increasing the intracompartmental pressure and leading to muscle and nerve ischemia. Therefore, we propose that the definition of Volkmann ischemic contracture should be maintained only for the acquired forms with an evident etiology and that Frieden’s classification scheme for ACC type VII needs to be reformulated.

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function although a slight hypotrophy of the limb was still present (Figure 2). On the basis of the suspected neurological involvement, the diagnosis of CVIC was initially considered, but after neurological evaluation the final diagnosis was ACC.

**Discussion**

A review of the literature (eTable in Supplement) evidenced that more accurate definitions of ACC and CVIC are needed because the rarity of these conditions has led to a lack of certain criteria for the differential diagnosis. According to Frieden,\(^3\) when ACC involves extremities without associated abnormalities, it should be included in type VII. Although this type of ACC more often shows a symmetrical distribution, in 8 cases it was found to affect only 1 limb.\(^2\) Of these, 2 were associated with skeletal abnormalities at birth and 1 with neurological impairment.\(^2\) Then, in 2012, the original classification was expanded, introducing 2 subtypes for type VII, in order to include the cases in which the skin defect was associated with the bony abnormalities.\(^7\) In such cases, the bone involvement could be related to the damage of perichondrial vessels induced by an external compression and resulting in the necrosis of the perichondrium, as well as the adjacent physeal cells.

The definition of CVIC was introduced for the first time in 1980 by plastic surgeons\(^6\) who applied the term to a congenital skin defect of the arm associated with ipsilateral nerve palsy. Since then, an additional 11 cases of CVIC have been reported in orthopedic surgery, plastic surgery, and dermatology journals, but the authors did not clarify how to differentiate the clinical manifestations from those of ACC.
Aplasia cutis congenita (ACC) is a skin condition that affects newborns, characterized by a partial or complete absence of skin tissue resulting in a cutaneous defect. The causes of ACC have included genetic factors, intrauterine trauma, cases in nerve palsy, bony defects, or even amputation. Other mesenchymal bands that surround fetal structures or adhere to the flexion of the ipsilateral wrist or, in more severe cases, hand paralysis. It can also be associated with muscular and neurological impairment; ACC does not present with any associated neurological involvement.

### Type Clinical Features

<table>
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<tr>
<th>Type</th>
<th>Clinical Features</th>
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<tr>
<td>I ACC of the scalp without multiple abnormalities: AD or sporadic</td>
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<tr>
<td>II ACC of the scalp with associated limb reduction abnormalities: AD</td>
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<tr>
<td>III ACC of the scalp with associated epidermal and organoid nevi, corneal opacities, and psychomotor retardation</td>
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<td>IV ACC of any site overlying embryologic malformations such as meningoencephaloceles or spinal dysraphia</td>
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<tr>
<td>V ACC of any site with associated fetor papyraceus or placental infarcts; mostly symmetrical and linear</td>
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<tr>
<td>VI ACC of the extremities associated with epidermolysis bullosa, usually on the extremities: inheritance depends on type of epidermolysis bullosa</td>
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<tr>
<td>VII ACC of the extremities without any other abnormality, or ACC of the extremities with bony abnormalities and/or neurological involvement</td>
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<tr>
<td>VIII ACC of the scalp caused by specific teratogens such as varicella or herpes simplex infections or in association with methimazole treatment</td>
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<tr>
<td>IX ACC associated with malformation syndromes such as trisomy 13 and ectodermal dysplasia</td>
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Abbreviation: AD, autosomal dominant.

*Adapted and revised from Frieden, 1986.*

Congenital Volkmann ischemic contracture, conversely, has been associated with prenatal conditions such as oligohydramnios, polyhydramnios, death of a co-twin, gestational diabetes mellitus, or delivery complications, but, as in ACC, external compression by amniotic bands or the umbilical cord seems to be the most viable hypothesis.

This article is partly focused on semantic issues because dermatology is well known for its “misnomers” and a rational nomenclature is mandatory for our discipline. The description of CVIC was introduced by orthopedists and plastic surgeons, who may not be accustomed to a rare dermatological disease such as ACC. The literature presents cases of ACC with palsy, without palsy, or with dysplasia of the radius, causing diagnostic confusion. Moreover, the pathogenesis of ACC overlaps with that of CVIC (eg, dead twin, amniotic bands).

In our opinion, CVIC should be considered a particular form of ACC in which an external noxa affects the forearm, increasing the intracompartamental pressure and leading to muscle and nerve ischemia. Therefore, we propose that the definition of Volkmann ischemic contracture should be maintained only for the acquired forms with an evident etiology and that the classification scheme of Frieden for ACC type VII might be updated as follows (Table):

Type VIIa. ACC of the extremities without any other abnormality;
Type VIIb. ACC of the extremities with bony abnormalities and/or neurological involvement.

### Conclusions

In the event of congenital ulceration of the forearm, we suggest performing a complete neurological examination; electromyography, if indicated; and radiography to rule out nerve palsy and/or bony abnormalities to achieve the correct diagnosis.

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**ARTICLE INFORMATION**

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**REFERENCES**


