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Case Report

Case Report: Newly Born with Multiple Articular Contractors. Differential Diagnosis of Multiple Congenital Arthrogryposis

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Abstract

Arthrogryposis comprises a heterogeneous group of different clinical entities that, both because of the clinic and because of the difficulty in an adequate therapeutic management, deserve a multidisciplinary approach.

The diagnosis is not confined as such to a specific clinical picture, since there are many alterations present at the time of birth that can simulate this entity.

We present the case of a 4-month-old girl with deformities in both lower limbs at birth. Both the presence of these, and the excessive hypotonia of the patient, led us to consider the possible etiology, including arthrogryposis.

After a follow-up and multidisciplinary treatment of the patient, the evolution of the same has been encouraging, considerably improving muscle tone and joint mobility, correcting the bilateral clubfoot, as well as the extension deformity of both knees. Even so, the difficulty of clinical management has been marked by hypotonia, respiratory problems, swallowing and reflux of the girl, for which she has needed hospitalization on several occasions. Currently, the etiology of the pathology is unknown since all the studies performed have been negative, leading only to the diagnosis of myopathy.

Keywords: *Multiple Congenital Arthrogryposis (AMC); Arthrogryposis; Myopathy*

Introduction

Multiple congenital arthrogryposis (AMC) is a group of disorders characterized by multiple joint contractures that affect the dorsal musculature, lower and upper extremities. The term arthrogryposis derives from the Greek arthron-articulation and grypos-corvo/hooked, means flexed joint. AMC is a progressive syndrome, not a disease, even for some authors it is classified as a symptom, present from birth [1].

The etiology is unknown, is attributed mainly to the lack of fetal movement, which may be due to neuropathies, muscle disorders, connective tissue abnormalities, as well as viral infections and restrictive intrauterine disorders such as structural alterations of the uterus, oligohydramnios, breech presentation or prematurity, causing, in this way, a paralysis of the fetus, causing alteration of the development and the extremities, producing scarring in the fetal musculature [2].

Only in 30% of cases is a genetic cause, the prevalence is estimated at 1 in 8000 RNV, no significant difference in presentation has been demonstrated in terms of gender.

Clinical Case

We present the case of a 11-month-old girl, who presented multiple contractures at birth, mainly of lower extremities, with extreme extension of knees and hyperflexion of hips, reaching to touch the anterior portion of the chest, also presented deformity in adducts and varus. of both feet (Figure 1).

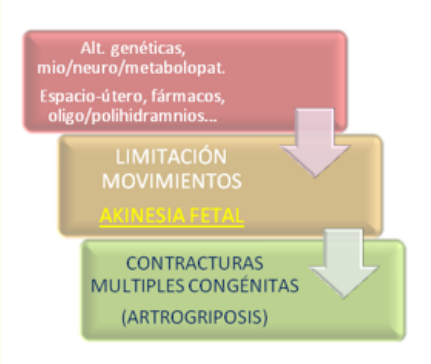

Figure 1

Discussion

The AMC was first described in 1841 by Otto, and in 1982 the term amyoplasia was introduced, which describes the most common type of arthrogryposis, which is characterized by multiple contractures, symmetrical position of all four limbs and replacement of skeletal muscle by tissue fibrous and fat. Up to now, at least nine different types of aphasia have been recognized [3].

Patients with AMC have multiple contractures, joint deformities and muscle abnormalities, which causes the extremities, sometimes, appear conical, fusiform or curved. Around 80% have contracture in the hip and 43% congenital hip dislocation, others have up to 90% of contractures in the knees, flexion contractures being more frequent, the feet are also affected, appearing up to 92% rigid equinovar foot. The proximal extremities are affected by around 60% and scoliosis is present in 35% of patients [2].

The diagnosis of AMC is made by exclusion, since there are 150 different disorders that manifest muscle contractures. The clinical history is essential, as well as the follow-up during pregnancy and childbirth [3].

The objective of the treatment is to obtain the best possible function of the patient, it must be multidisciplinary and be established as early as possible, including management by trauma, pediatrics, rehabilitation and neurology. Physiotherapy is essential for the development of a better function of the child, as well as corrective surgeries of deformities in fixed contractures [1].

In general, the development of the intellect is not usually affected and the prognosis in terms of the patient's function depends on the early start of the treatment. As factors of good prognosis, hip flexion contracture of less than 20º and flexion knee contractures of less than 15º can be considered, however studies have concluded that long-term ambulation does not correlate with contractures at birth [4].

Conclusion

For some authors, arthrogryposis is considered a "sign" of an extensive etiology, produced by extrinsic, intrinsic, genetic or environmental factors; all having as their central axis the fetal akinesia (Figure 1).

In our case, the possible triggering factors could be a fetal breech presentation and an external version, performed one month prior to birth. The premature loss of fetal well-being, contractures, cyanosis, absence of suction reflexes and the alteration of the patient's muscle tone, did not lead us to consider CMA as a differential diagnosis, despite this, the diagnosis and filiation of the pathology. of our patient, is not yet known.

However, despite the complications presented in our clinical case, we can say that the evolution of the patient has been positive. Being treated initially with Pavlik harness and orthosis for the forefoot adduct, it is currently able to maintain standing and improve dependence for acts such as swallowing. However, they are patients with a high demand for care and a great need for care, therefore the management and early treatment of them is imperative (Figure 2-4).

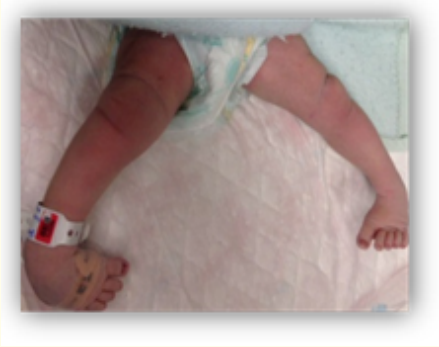

Figure 2

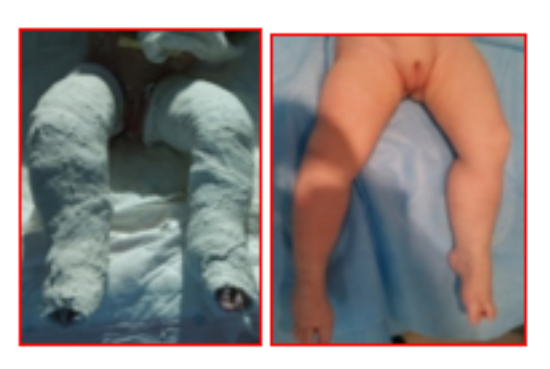

Figure 3

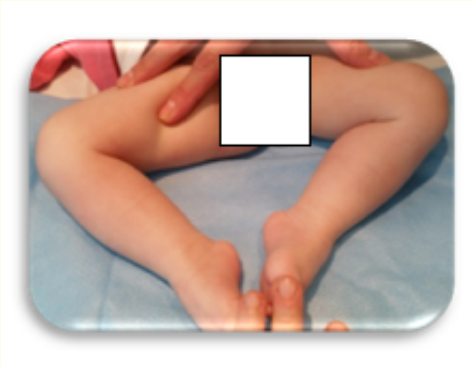

Figure 4

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