Early Splinting Intervention and Active Motion Achievement In Child with Amyoplasia - A Case Report

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INTRODUCTION

Amyoplasia is the most common arthrogrypotic disorder, characterized by contractures of multiple joints and symmetrical positioning of the limbs. In most patients the upper extremities present adduction and internal rotation of the shoulders, extension contracture of the elbows, pronation of the forearms, flexion of the wrist, adduction of the thumb and flexion of the PIP joints. Most authors refer to stiffness as extra articular and recommend passive range motion exercises (PROM) starting early in life; the importance of daily therapy; home program and splinting at night to maintain the achieved position. But most agree that therapy, by itself, rarely achieves significant improvement.

PURPOSE: The purpose of this case report is to show the result of early intervention in a child diagnosed with Amyoplasia, treated with a splinting program that included day and night use, early developmental stimulation and family orientation.

CASE REPORT:

S.A.S, 30 months, arthrogryposis detected before birth by Ultrasound. She was evaluated and identified the characteristic contractures of Amyoplasia in the lower and upper extremities. Hand Therapy treatment started on the first week of life. At first, positioning splints were made for the hands, PROM to all upper and lower extremities were initiated and parents oriented to perform at home. The splints were remolded every week.

At three months of age, S started to present the elbow extension pattern and a 30° flexion dorsal elbow splint was fabricated for both elbows to be worn during the day, for short periods, in order to control the extension movement. These elbow splints had straps at the arm, but the forearm was free to move. Within two weeks, light active elbow flexion was observed and an early developmental stimulation program started.

At the present age, S has active flexion of the elbows, free movements of the shoulder, active extension of the wrists and the thumb is abducted. She is also able to bring her hands actively to her mouth and although, the upper limb motion is weak, she is able to hold the walking aid and make progress on deambulation, manipulates toys of any sizes reaching, holding and throwing them, combs her hair and eats independently.

CONCLUSION: Authors believe that, during the elbow extension, co-contractions occur, otherwise elbow would hyperextend, not only the extensor is activated, some activity of weak flexors are present. Inhibiting extension, contraction of the flexors is more effective and active movement stimulation showed to be possible.

A hand therapist while treating and splinting a child with Amyoplasia must observe PROM, positioning and also the effects of them during muscle activity. This case report shows that the approach in therapy must be global and challenging, since active elbow flexion in this pathology is not described and it suggests that a serial of cases should be performed to sustain the hypothesis here presented.