

## Case Report

# Open quadricepsplasty in hyperextension deformity of knee in children with arthrogryposis

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### ABSTRACT

Arthrogryposis includes heterogeneous disorders, characterized by congenital contractures of multiple joints. Knee involvement is very common (38–90 % of patients with amyoplasia) ranging from soft-tissue contractures (in flexion or hyperextension) to subluxation and dislocation. Children who present late will require surgery involving quadricepsplasty or lengthening of the contracted quadriceps muscle. Curtis and Fisher describe an open V-Y lengthening of the quadriceps femoris with post-operative immobilization in 30-45 ° of flexion. The main issue of this technique was limited degree of flexion that can be achieved, and the fibrous replacement of the quadriceps muscle. This is a neglected case of hyperextension deformity and congenital dislocation of knee in children with arthrogryposis resistant to conservative management and successfully treated by open quadricepsplasty.

**Keywords:** Arthrogryposis, Congenital subluxation of knee, Curtis and fisher technique, Contracted quadriceps muscle, Hyperextension deformity of knee

### INTRODUCTION

Arthrogryposis multiplex congenita (AMC) is a nonprogressive condition characterized by the presence of multiple joint contractures at birth, and occurring in approximately one in 10000 live births.<sup>1-5</sup> In a simplistic and practical way, disorders with arthrogryposis have been subdivided into three large groups: amyoplasia, distal arthrogryposes, and everything else.<sup>6</sup> The most common recognizable form of arthrogryposis (representing one-third of all cases) is known as “amyoplasia”, a sporadic disorder, characterized by symmetrical limb involvement, some truncal sparing, normal to above-average intelligence, and frequently a midfacial hemangioma.<sup>3,4,7</sup> treatment is challenging because of the frequency of contracture recurrences.<sup>1</sup> Knee involvement is very common (38-90 % of patients with amyoplasia) ranging from soft-tissue contractures (in flexion or hyperextension) to subluxation and

dislocation. Children who present late will require surgery involving quadricepsplasty or lengthening of the contracted quadriceps muscle. We are reporting a case of congenital hyperextension of the knee in children with arthrogryposis treated by open quadricepsplasty.

### CASE REPORT

A 2-years-old female was admitted to the hospital due to deformities in her lower legs. She has been diagnosed with arthrogryposis multiple congenita since she was born, with bilateral CTEV, congenital subluxation of the right knee (Figure 1), and congenital right quadriceps contracture.

The hyperextension of the knee found to be resistant to conservative correction. We performed open quadricepsplasty using Curtis and Fisher technique, with long anterior incision, quadriceps mechanism then

divided by inverted V incision superior to the patella (Figure 2) and put post operative immobilization in 30° of knee flexion in above knee plaster of paris cast (Figure 3).



**Figure 1: Knee radiograph before surgery showed congenital subluxation of right knee.**



**Figure 2: V-Y Quadricepsplasty with long anterior incision.**



**Figure 3: Post-operative immobilization.**

On the last follow up, the active and passive ranges of motion (ROM) of the knee were measured with the help of a goniometer. The quadriceps power was tested by manual muscle testing and graded on the Medical Research Council (MRC) scale (Figure 4).

Three weeks after surgery, patient recovered well without any complication. The cast was opened, the operation wound was good and the post operative passive and active ROM was 45° which represented an improvement of 45° from the pre-operative ROM of 0°. The post operative quadriceps power was four, no difference from pre operative power.



**Figure 4: Three weeks follow up with the knee in 45° of flexion.**

## DISCUSSION

The treatment of the knee hyperextension contracture in AMC has been recommended to start early. Passive mobilization, serial casting and braces have been recommended. For this early treatment there have been good results especially in young children. The difficulties arise in cases of failure of these non-operative maneuvers or when the deformity presents in a severe and fixed form in older children. In such cases of fixed hyperextension surgical treatment with the quadricepsplasty is indicated.<sup>5,8-10</sup> There is some disagreement as to the surgical technique (percutaneous or open) that should be used.<sup>11,13</sup> In the open technique (recommended by most authors) either a Z-plasty or an inverted V-Y plasty of the quadriceps tendon is performed through an anterior approach.<sup>9,11,12</sup>

Curtis and Fisher describe an open V-Y lengthening of the quadriceps femoris with long anterior incision, we use this technique to achieve sufficient amount of quadriceps lengthening.<sup>9</sup> According to Staheli, the best position for the knee joint is in approximately 15° of flexion for both standing and sitting, we try to achieve more than this amount (post operative immobilization in 30° of flexion) without losing muscle power.<sup>5</sup>

Post operative wound was good, although the scar was quite long. Active extension of the knee was possible with no decrease in quadriceps muscle power. The main issue of this technique was limited degree of flexion that can be achieved, and the fibrous replacement of the quadriceps muscle. Curtis and Fisher reported seven from fifteen knees were considered to have good or excellent functional result.<sup>9</sup> Our case reported good result in active and passive ROM and good muscle power, but the patient still not allowed to walk and we can't evaluate the functional outcome and the gait pattern. Long term follow up will be evaluate for then future result.

## CONCLUSION

Open quadricepsplasty was effective in achieving adequate and functional correction in hyperextension deformity of knee in children with arthrogryposis.

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