Arthrogryposis Multiplex Congenita (AMC): A case report

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Introduction

Arthrogryposis multiplex congenita (AMC) is a rare congenital disorder that is characterized by multiple joint contractures and can include muscle weakness and fibrosis. It is a non-progressive disease. The disease derives its name from Greek; literally meaning 'curved or hooked joints.

In some cases, few joints may be affected and may have a nearly full range of motion. In the most common type of arthrogryposis, called amyoplasia, hands, wrists, elbows, shoulders, hips, feet and knees are affected. In the most severe types, nearly every joint is involved, including the jaw and back. Frequently, the contractures are accompanied by muscle weakness, which further limits movement. AMC is typically symmetrical and involves all four extremities with some variation seen. [1, 2, 3]

Case report

Here reported case is of 7 years old male child born with blood group A +ve and suffering from AMC. At the time of birth patient had bilateral Congenital Dislocation of Knee (CDK) and bilateral Congenital Talipes Equino Varus (CTEV). Child was not able to stand or walk due to deformities. Patient has been operated for the same seven times with serial plaster casts in his 7 years of life.

At the age of 5 years on examination patient had CTEV and bilateral CDK with 40^0 Fixed Flexion Deformity (FFD) of Right knee, recurvatum with 20^0 valgus of Left knee and bilateral radial head dislocation with restricted supination. Patient was operated for Left quadriceps-plasty, reduction of knee joint, bilateral Tendo Achilles (TA) Tenotomy, Plantar fasciotomy and Tibialis Posterior release. After surgery patient was advised for strict limb elevation and active toes movements followed by physiotherapy.

At the age of 6 years on examination patient had recurred CTEV and ended up in fixed deformity including varus, adduction, cavus and equines and patient was operated for bilateral dorsal wedge resection of foot & k-wire fixation, TA tenotomy & posterior capsulotomy of ankle and subtalar joints bilaterally. Four months after surgery, on clinical examination residual knee FFD of 10^0 of right knee and resistant left foot deformity for correction to neutral position.

At the time of presentation to physiotherapy clinic after last surgery for physiotherapy patient had reduced muscle power on both lower limbs. By manual muscle test described by Daniel muscle power was reduced in all muscles of lower limbs, but in majority of muscles power was found > 3 out of 5.[4] On girth measurement, wasting of was found on left side as compared to right side. Amount of wasting found was 0.5 cm and 1 cm respectively in thigh and leg. Limb length shortening was found on right side as compared to left side. Amount of shortening found was 6 cm and 2 cm in above knee and below knee measurement.[5]

Patient before surgeries

Patient after surgery (Supine)

Patient after surgery
(Walking with rollator)

Physiotherapy treatment was directed towards maintenance of range of motion in all joints of bilateral lower limbs, strengthening of weak muscles of bilateral lower limbs as well as gait training with bilateral ankle foot orthosis and knee extension brace and with rollator. Bilateral upper limbs were only given home exercises and patient had become habitual with the use of both upper limbs even with restricted supination range of motion. Along with this physical treatment, psychological counseling of patient's father about regular follow-up to orthopedic surgeon and consistency in visit of physiotherapy department without fail was done.

Discussion

The patient selected for the study was not able to walk before surgery as the above mentioned muscles were very short and not allowing movement in full range of motion. Due to short muscles since birth there was also development of above mentioned deformities. If child would have been untreated, he might not have been able to stand on his own. Preoperative physiotherapy, surgeries with serial plaster casts and postoperative physiotherapy made the child walking with the help of assistive devices like splints and mobility aids like rollator. There are very few researches available for role of physiotherapy in arthrogryposis multiple congenita and other related congenital abnormalities. So more numbers of researches are needed to be done with long term duration. This can also help for creating awareness about changing common belief that for this type of child, only maintenance of condition and preventing complication is not sufficient. This type of patient can be given improved quality of life even though the condition itself is not curable.

Conclusion

Arthrogryposis multiple congenita is rare congenital disorder. It is not curable but by multiple corrective surgeries and physiotherapy treatment, child can be made independent as much as possible. Before surgery, patient should be assessed for manual muscle testing to decide the possible outcomes of surgery as muscle power reduces after surgery. After surgery, if physiotherapy treatment given properly, child can live with better quality of life later on.

References

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