

Orthotic Management of Patient with Arthrogryposis Multiplex Congenita - A Case Study

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Abstract: *Arthrogryposis Multiplex Congenita (AMC) is a syndrome characterized by non-progressive multiple congenital joint contractures. The etiology of disease is multi factorial. It is most commonly suspected from absent fetal movements and genetic defects. Great number of pathological processes causing immobilization of limbs of a fetus during or shortly after the embryonic formation of joints may result in AMC. Rehabilitation should be started in early stage for functional independence. Patient with AMC may have lower extremity contractures or upper extremity contractures or both. Lower extremity contractures and muscle weakness are common in AMC. Consequences of muscle weakness and joint contractures in the lower limb influences walking ability of the person. Severity of contractures and pattern of deformities will change from person to person depending on the prenatal conditions. As there is no fix pattern of deformities designing of orthoses is very crucial part and it should be done by keeping in view individual person. Before designing any orthotic design, basic consideration should be given to the available range of joint movement, severity of contractures, age of the patient and their personal and professional requirements. The aim of this study was to develop the custom-made orthotic design for AMC patient. Though it is an external support hence called as Orthoses but most of the times to compensate for the limb length shortening prosthetic components are also used. As it is a combination of orthoses and prostheses hence also referred as Extension Ortho-Prosthesis.*

Keywords: Arthrogryposis Multiplex Congenita, available range of joint movement, severity of contractures, custom-made orthosis, Extension Orthoprosthesis

1. Introduction

Arthrogryposis Multiplex Congenita (AMC) is a syndrome characterized by non-progressive multiple congenital joint contractures. Arthrogryposis Multiplex Congenita is a common name caused by many different syndromes rather than a single syndrome. The joints of the lower extremities are more affected than the upper extremities. In addition, the jaws and back may also be affected in individuals with AMC. There are two main types of the disease. Amyoplasia is the most common (40%) and classical type^[5]. Second type is Distal

Arthrogryposis. Both of these types are characterized by contractures of various joints. The symptoms of AMC are present at birth however; specific symptoms and physical findings can differ greatly in range and severity from one person to another.

In this paper attempt is made to designed custom made lower limb orthosis for AMC patient who is using the orthosis since childhood and now expecting better design which will help to enhance the quality of personal and professional life.



In children with AMC the ability to walk depends on the extent of joint range of motion, in particular the hip joint, knee joint as well as in the ankle and feet, with the possibility of a plantigrade foot position. Knee joint involvement has been reported in 70% most of those are

flexion contracture. In children with Amyoplasia hip deformities are common and ranges from soft tissue contracture to hip dislocation^[3, 4]. Foot deformities are frequently observed with equinovarus and adductus foot being the most common^[4, 5]. In children with

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Amyoplasia, muscle weakness primarily in the hip and knee extensor muscles, influence walking ability [6]. Muscle weakness is reported to be more of an influence than contracture severity on walking ability. Equinovarus is often associated with plantar flexor muscle weakness [6].

The aim of this article was to design and test the light weight and cosmetically good Extension Orthoprosthesis for AMC patient that is designed to provide efficient gait pattern.

This research was reviewed and approved by the institutional Ethics Committee. Informed consent was obtained from participant.

2. Literature Survey

The correct diagnosis and the initiation of appropriate rehabilitation programme in terms of multidisciplinary approach are essential for the treatment. Splints or serial casting, passive stretching and range of motion exercises are effective in the treatment of contracture. Early surgical release of contracted tissue of joints which constitutes the basis of the problem is crucial for the prevention of the deformity [1]. As surgery has very limited role in the rehabilitation of AMC mostly these patients are dependent on orthosis throughout the life. Pattern of deformity will change from person to person hence while treating AMC patient it is important to customize the orthosis depending on the available range of joint movements, severity of contractures, age of the patient, professional demand and patient's expectation.

Due to the instability at the ankle joint, these children generally unable to stand still without external support resulting in increased knee flexion. The patient with a longitudinal deficiency may require an Extension Orthoprosthesis. Extension Orthoprosthesis is the combination of orthotic and prosthetic component. Prosthesis replaces a part of extremities and an orthosis stabilizes the existing extremity. The Extension Orthoprosthesis compensates the longitudinal malformations, have a supporting effect, control progression of contractures and compensate for the shortening. This may be combined with surgical reconstruction.

3. Problem Definition

Patient enrolled for the study was 22 year old female who is working in information telecommunication sector. She is a case of AMC with left side lower extremity involvement having knee flexion deformity of 35-degree, hip flexion deformity of 10-degree, ankle Equinus deformity of 20-degree along with foot inversion and internal rotation at hip. She was using an orthosis since childhood which is made up of combination of metal and plastic.

Problems in the present designed are:

1. Heavy in weight.
2. Not total contact hence provides less stability.

3. Anatomical alignment of design not fulfilling biomechanical requirements
4. Not able to control internal rotation of the foot as footplate was fixed.
5. Energy expenditure while walking was more.
6. Cosmetically not good.



4. Methodology / Approach

Considering the problems with present designed it was aimed to fabricate lightweight, total contact, modular Extension Orthoprosthesis with minimum components which can give better energy saving gait pattern than previous design.

Patient's casting was done in possible corrected position. As per patient measurement a lightweight orthoprosthesis was designed. This design consists of a single piece total contact plastic molded above knee shell, modified ankle adaptor and Polyurethane foam foot piece. Polyurethane foam foot helps to reduce the weight of the extension orthoprosthesis. Modified ankle adaptor is made up of mild steel material. Aluminum pipe is attached to the steel plate and distally with the washer by means of electric welding. This modification helps to provide internal or external rotation of the foot as per the need.

Main consideration was given to make extension orthoprosthesis lightweight, total contact and cosmetically acceptable. Internal rotation of hip caused difficulty in dynamic alignment of orthoprosthesis which was solved by using modified adaptor.





5. Results & Discussion

Arthrogryposis Multiplex Congenita is diagnosed prenatally by ultrasound or during birth. In this case family history was normal. Diagnosis had been made just after the birth due to characteristic multiple joint contractures.

Total contact design helped in better pressure distribution which increases the comfort. There is no extra metallic bar and polyurethane foam foot combination dramatically reduced the weight and helped to reduce the energy expenditure while walking. Facility of foot rotation by means of modified ankle adaptor helped to compensate internal rotation of the hip joint during walking which had improved the gait pattern. Provision to adjust external rotation of foot had helped to increase walking base of support. There was no tendency for knee flexion in swing due to total contact which increases the stability. It was cosmetically better hence better accepted by the patient.

6. Conclusion

Lower limb contractures and muscle weakness are common in Arthrogryposis Multiplex Congenita. Stretching and passive range of motion exercises for contractures; active resistive exercises for extremity strengthening, balance, proprioception and walking exercises are integral part of rehabilitation of person with Arthrogryposis Multiplex Congenita. Splinting of lower limb is also very important because person is dependent on the splint throughout the life for ambulation. Appropriate design of orthoses is very important to enhance the quality of life. As pattern of deformities will change from person-to-person custom design of orthoses needs to be developing depending on the individual requirements and availability of modern components.

7. Future Scope

When deformity pattern is not standard there is wide scope for modifications in the orthotic design. Design in this study was fabricated by using locally available

materials and technology. As more research takes place in material technology extension orthoprosthesis can be fabricated using more light weight material and enhanced cosmetic appearance which will aid in the better gait pattern of the patient.

References

- [1] Eriksson M, Gutierrez-Farewik E, Brostrom E, Bartonek A (2010): Gait in children with Arthrogryposis Multiplex Congenita. *J Child Orthop* 4: 21-31
- [2] Florence J (1977) The orthotic management of Arthrogryposis. *Journal of Prosthetic and Orthotic International* 1: 111-113
- [3] Sells JM, Jaffe KM, Hall JG (1996) Amyoplasia, the most common type of Arthrogryposis: the potential good outcome. *Paediatrics* 97: 225-231
- [4] Guider KJ, Drennan JC (1985) Foot and ankle deformities in Arthrogryposis multiplex congenita. *Clinical Orthopaedics* 194: 93-98
- [5] Hoffer MM, Swank S, Eastman F, Clark D, Teitge R (1983) Ambulation in severe Arthrogryposis. *Journal of Paediatrics Orthopaedics* 3: 293-296
- [6] Staheli LT (1998) Lower extremity management in Arthrogryposis. A textbook of Atlas. Cambridge University press 55-73.
- [7] Bevan WP, Hall JG, Bamshad M, Staheli LT, Jaffe KM, Song K (2007) Arthrogryposis multiplex congenita (Amyoplasia): an orthopaedic perspective. *Journal of Paediatrics Orthopaedics* 27: 594-600
- [8] Hall JG (1997) Arthrogryposis multiplex congenita: Etiology, genetics, classification, diagnostic approach and general aspects. *Journal of Paediatrics Orthopaedics* B6: 159-166
- [9] Thompson GH, Bilenker RM (1985) Comprehensive management of Arthrogryposis multiplex congenita. *Clinical Orthopaedics* 194: 6-14
- [10] M. Bamshad, A. E. Van Heest and D. Pleasure, "Arthrogryposis: a review and update," *The Journal of Bone and Joint Surgery-American Volume*, vol.91, No.4, pp.40-46, 2009.

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