

Case Reporte

A Case Report of Bilateral Upper Extremities Arthrogryposis

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Arthrogriposis is a disorder in which two or more joints in more than one limb of body, place in a stable position and malformation status that usually is the result of joints' immobilization during fetal period. Problems resulting from this congenital deficit can occur in difference places, in upper and lower limbs, trunk, and even in face. It occurs in 3 of 1000 births, in two-third of these patients, all four limbs suffer and one-third of patients suffer in lower limbs and it rarely occurs in upper limbs. In this case affected children and their parents try different treatment such as surgical and non-surgical treatments which are of argument and controversial even among specialists of these treatments. In this paper, a 3-year old boy has been introduced who suffers from congenital deficit with developmental disorder in upper extremity bones and the absence of flexor muscles group and extensor of wrist. The boy was sent to Bahar Rehabilitation Center for occupational therapy and rehabilitation services by physician of rehabilitation clinic in Tehran.

Key words: Arthrogryposis, Upper extremity, Rehabilitation

Introduction

Arthrogryposis Multiple Congenital is one of the congenital defects that has been observed during birth, the exact cause of this has not been still known but variety of causes like decrease of foetus movement, damage to anterior horn of spinal cord, genetic reasons, lack of amniotic liquid, unnatural growth of connective tissue, viral or bacterial infections and endocrine disorders etc. can be effective in this disorder (1, 2).

Usually the joints are involved bilaterally. In general, this disorder can be divided into two groups: neurological and non-neurological. First group includes central and peripheral nervous system disorders and the second group includes rheumatologic disorders and movement limitations (2).

Classic arthrogryposis or amyopalsia was first suggested by Shlordane in 1932. This disorder is due to lack of muscles groups and occurs because of muscles replacement which happens by fibrous or fat tissue. The most prevail type of this disorder involves both lower and upper limb joints and rarely

involves upper extremity isolatedly; it occurs 1 to 5 in 10000 births (3).

Clinical manifestations in upper extremity has more affected by elbow joint that could be in flexion and extension positions. In the first case because of flexion in child's elbow joint, he/she can use his/her hand to reach his/her mouth and face for hygiene or eating, but in the second case which is more important, elbow extends, arm bone and forearm rotate internally and wrist is inflexed which is called "waiter's hand" (4, 5).

The only advantage of this shape of the hand is in hygiene of genital area. In this case the child is unable to bend elbow joint to eat or brush his teeth. These children usually have delayed movement growth such as crawling, and getting up from ground and walking. Usually they move from one place to another place with the help of upper extremity when their buttock is on the ground, because in this condition their dorsal of the hand is on the ground, the extent of motion increases and also increases the lumbar lordosis. Usually in these children upper extremity is shorter than should be in normal case. Treatment of these children depends on the present

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circumstances including the range of motion, situation and the power of upper extremity muscles. The final goal in treatment of these patients is to help them being able to eat and do their face and genital area hygiene (8-9).

The main current surgical option for these children is elbow capsulotomy and triceps release. This operation is usually for bending elbow joint in a passive way and if we want to bend the elbow in an active way, we must transfer one of the triceps, pectoralis major, wrist and fingers flexors and latissimus dorsi in addition to what has been mentioned. In order to transfer of each of the mentioned parts, there are different ideas among muscle surgeons. Such transfers also depend on rang of motions of the shoulder and elbow and wrist and the most crucial item is the power of the muscle which has been transferred (9-11).

In treatment of these children, it is necessary to know that treatment of each person is unique and is based on the extent of limitation of upper extremity and the level of independence in basic activities that is vital for the person.

All rehabilitation team members including physicians, therapists, orthopedic surgeons, psychologists and so on must be well informed about their crucial role in patient management and counseling and explaining the treatment options and each option advantages or disadvantages.

The involved team including psychologist, occupational therapist, physiotherapist, orthopedic surgeons, technical orthopedic must work together and explain to parents the ways of treatment. Rehabilitation team has a key role before and after the surgery.

In order to improve the range of motion and muscle reinforcement, occupational therapist and physiotherapist should use support traction exercise, manipulation, warming, serial casting and hydrotherapy prior to surgery. To increase function and maintain the range of motion, technical orthopedics should make suitable splint or orthosis considering the patient conditions as well. Occupational therapist can also have a very important role in designing and using equipments to make daily activity of patients independent. After the operation, rehabilitations are very important. Orthosis-prosthesis prescriptions must be accompanied with a complete assessment and should be continued to six months after operation. By active/passive movement of uncasted joints we must stop the stiffness of joints during non-movement

period. Muscle exercise should be the priority of rehabilitation exercises, if muscle transfer happens (12-13).

Because this disease is rare and therapists have little information about it, such patients have many problems (in addition to psychosomatic and physical disorders) the different kinds of treatment for these patients, cause stress and confusion for their parents. Rehabilitation treatment is a crucial and effective part of treatment both before and after the surgery. Thus, knowing the disease and the role of therapists are the most important factors in the treatment of affected persons.

Case report

The patient was a 3-year old boy whom was recognized as having the bilateral upper extremity due to arthrogryposis. To improve the range of motion and to reinforce the daily activity training by therapist recommendation, he came to Bahar Rehabilitation Clinic in Tehran. The parents were not related and they did not have any history of such disease in their family. Mother was 38 years old when she got pregnant and the child was born through cesarean section. The birth time weighed and height was 3/5 kg and 50 cm, respectively and head circumference was normal. Radiography showed severe mal-development on elbow and wrist joints, arm bone and forearm, when he was 2.5 years old. Because the wrist of infant was bent, steps of motion development like grasping things had very slow development but other steps of motion were almost in time, as the parents explained, he could control his head and neck when he was 4 months old, he could sit when he was 7 months old, he was able to move by sitting on his buttock when he was 9 months old, and he could stand up straight from sitting when he was 13 months. Other steps of motion development like crawling were not done because of disorder in toddler. Because the toddler was able to bring up his hand from his shoulder and had good muscle strength, all of the rehabilitation treatments were related to increase range of elbow flexibility. Although the wrist flexibility would help him eat by himself, but for holding things he needs to bring up his wrist. Therapist should not forget to straighten his wrist. The child was able to put the pieces of food in his mouth with the assist of the other hand, when he was 3\5 years old and with the increase of range of motions of elbow of his right hand, was able to do most of his personal activities when he was 4. Some surgeons believed that

operation must be done in younger ages and some others recommended postponing the surgery. After bone growth and joint plates completed, some believed that one hand should be bent and fixed and then operation can be done on them, and some believed that one hand should be bent and not moved. Therefore one hand performs the needs of upper half, and the other hand stays straight to do the needs of the lower half of the body, and also in the

case of muscle surgery, some suggest pectoralis major muscle and some others surgeons suggest triceps release. But rehabilitation treatment have been done with emphasis on practices of muscles to improve their strength, improvement of range of motion, prevention of joints deformation, the mental support of parents and infant, and teaching the parents to teach their infant how to be independent.

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