Introduction

The first western report of an arthrogrypotic infant is attributed to Adolf Wilhelm Otto in 1841.He described a stillborn infant with multiple congenital joint contractures as a ‘‘human monster with inwardly curved extremities.’’ Rosenkranz in 1905 have been the first to use the term ‘‘arthrogryposis’’ in a descriptive way, meaning only ‘‘curved joint.’’ In 1923 Stern gave these disorders the name arthrogryposis multiplex congenita ***(Ezaki, 2010).***

Arthrogryposis or arthrogryposis multiplex congenita (AMC) is a generic term used to describe the presence of multiple congenital contractures. The word arthrogryposis, arthro, joint, gryp, curved, literally means curved joint (implying that it is fixed or stuck in the curved position). Thus, arthrogryposis multiplex congenita means curved (fixed) joints in many (multiple) areas of the body, which are present at birth (congenita) ***(Staheli et al., 2008).***

Amyoplasia is the most common of the arthrogrypotic conditions and is the one with which orthopaedic surgeons are most familiar. Amyoplasia is nonprogressive, nonhereditary, and not associated with major organ system disorders. Children with amyoplasia can be expected to have normal sensation, normal intelligence, and a normal life expectancy ***(Ezaki, 2010).***

Arthrogryposis is an uncommon problem.Because there are many causes, correct diagnosis is important to predict the natural history and determine appropriate treatment. Inconsistent terminology has caused confusion about both diagnosis and treatment ***(Bernstein, 2002).***

The upper extremities tend to be in classic posture of internal rotation at the shoulders, extension of the elbows, and flexion at the wrists. The lower extremities show rigid flexion and internal rotation at the hips, stiffness in the knees, and clubfeet. In the “classic” case of arthrogryposis, hands, wrists, elbows, shoulders, hips, feet and knees are affected. In more severe cases, nearly every body joint may be involved, including the jaw and the back. Frequently, the contractures are accompanied by muscle weakness, which further limits movement ***(Alfonso et al., 2000).***

The causes of arthrogrypotic syndromes are largely unknown, but are presumed to be multifactorial. Potential etiologies have been grouped by Hall into neuropathic abnormality (brain, spine, or peripheral nerve), abnormality of muscle structure of function (muscular dystrophies, mitochondrial abnormalities), abnormality of connective tissue (diastrophic dysplasia, DA), intrauterine space limitation (increased prevalence in twins), maternal diseases (multiple sclerosis, myasthenia gravis, trauma), and impaired intrauterine fetal vascularity (impaired normal development of nerves, anterior horn cell death) ***(Bevan et al., 2007).***

Diagnosis is made by ruling out other causes. Muscle biopsies, blood tests, and clinical findings help rule out other possible disorders and provide evidence for arthrogryposis. AMC can sometimes be diagnosed during pregnancy. Ultrasounds at approximately 20 weeks gestation may show an abnormal position of joints or lack of joint movement. Alternatively, the diagnosis can be made on the basis of clinical symptoms and findings. The specific subtype can often be pinpointed with radiographs. Sometimes, an electrical nerve or muscle conduction study is necessary. CT scan or MRI can identify any central nervous system abnormalities or myopathic forms and may provide important information ***(Zhao et al., 2007).***

Treatment of arthrogryposis is often challenging and should not be undertaken by any one doctor, but by a multi-disciplinary team, including the pediatrician, neurologist, orthopedic surgeon, geneticist, physical and occupational therapist. The aim of treatment is to improve function. Intervention includes therapy, splinting, and surgical procedures ***(Hall, 2007***