Arthrogryposis Education Page

What is Arthrogryposis?

Arthrogryposis (Arthrogryposis Multiplex Congenita) is a non-progressive physical disability characterized by the presence of multiple fixed joints throughout the body at birth. Limited movement of the fetus in the womb results in the formation of extra connective tissue, which becomes fixed around the joints. The tendons, which enable motion, either do not form or do not stretch out completely. This disrupts normal muscle development and limits the range of motion around the affected joints. Arthrogryposis typically affects joints of the hands, feet, shoulders, hips, knees, wrists and elbows and in some cases various muscles throughout the body.

How often does Arthrogryposis occur?

Arthrogryposis is estimated to occur in 1 out of every 3,000 children born. The general incidence of a congenital contracture (clubbed foot/hand, dislocated hips, etc.) is 1/200 children.

What are the most common forms of Arthrogryposis?

Arthrogryposis is an umbrella diagnosis describing a number of conditions in which multiple contractures (joints around which movement is impaired) are present. There are over 150 variations of Arthrogryposis. There are three generally accepted categories of Arthrogryposis:

1) conditions affecting only the arms and legs
2) conditions affecting the limbs and other areas of the body
3) conditions affecting the limbs and central nervous system

60% of people diagnosed with Arthrogryposis are affected in all four limbs.
25% of people diagnosed with Arthrogryposis are only affected in their legs.
15% of people diagnosed with Arthrogryposis are only affected in their arms.

Amyoplasia, characterized by a lack of or abnormal muscular growth, is the most common form of Arthrogryposis. It is estimated that approximately 33% of people diagnosed with Arthrogryposis have Amyoplasia.

Distal Arthrogryposis is characterized by the presence of contractures in the hands and feet. Distal Arthrogryposis results from genetic mutations. There are several sub-types of Distal Arthrogryposis, distinguished by their associated physical characteristics. Type 1 Distal Arthrogryposis typically involves overlapping fingers, clenched hands, finger contractures and clubbed feet.

Common Characteristics and Complications Associated with Arthrogryposis:

Clubbed Feet/Hands	Internally rotated shoulders & limbs
Dislocated hips& joints	Muscle(group) absence, malformation, atrophy
Scoliosis	Webbed &/or Dimpled Skin
Hernias	Respiratory Problems & Aspiration of Fluids
Facial Asymmetry	Intestine, kidney, bladder and genital malformation
Facial Birthmarks	Decreased Muscle Mass

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What causes Arthrogryposis?

While there has been no single, conclusive cause attributed to the occurrence of Arthrogryposis, the general consensus is that anything that interferes with the normal in-utero or intrauterine growth or movement of the fetus can cause Arthrogryposis. These are thought to include: decreased space in the womb for the fetus to move around, maternal fever, viral infections, and/or environmental factors. The severity of Arthrogryposis is thought to be related to how early in the pregnancy the reduction in movement begins. Shortly after birth, the diagnosis of Arthrogryposis is confirmed w/ a physical examination, the collection of medical history information, x-rays and diagnostic tests including blood tests and muscle biopsies.

Is there any treatment for Arthrogryposis?

Specific courses of treatment vary, depending on the diagnosis and level of severity of Arthrogryposis. Typically, a continuous regimen of surgery, casting, splinting and physical and occupational therapy is used to increase the child's range of motion and functionality. Surgery is used to correct certain deformities that can not be improved by splinting and casting. Soft tissue operations include soft tissue releases, in which the muscle/tendon, ligament, nerve or artery is cut from its fixed position and in some procedures manually lengthened, and tendon transfers. These are performed on younger patients and to correct less severe deformities. Osteotomy (bone procedures) are performed on older children and to improve more severe deformities. These include reductions (the correction of dislocated joints, commonly the hips), the removal of wedges of bone which are fused and talectomy (removal of the talus bone in the foot).

Non-surgical methods of treatment include: serial positioning, casting, splints, manual stretching and physical and occupational therapy. Serial positioning is the use of foam wedges strategically placed to help align an infant's body. The angles of the wedges are gradually reduced to improve positioning and the infant's range of motion. Casting, which can be done serially or after surgical procedures, and splints are used to maintain the positioning, range of motion, stability and flexibility achieved through surgery, manual stretching and occupational and physical therapy. Manual stretching is done both in therapy and during home exercises for flexibility and muscle lengthening. Physical and Occupational Therapy services include: increasing the range of motion, splinting, the introduction and use of adaptive equipment, mobility training and self-care skills.