



Arthrogryposis

Symptoms or Behaviors

- Normal Intelligence
- Normal speech
- Intact sensation
- Muscle weakness
- Internal rotation deformity of the shoulder
- Extension and pronation deformity of the elbow
- Volar and ulnar deformity of the wrist
- Finger in fixed flexion and thumb-in-palm deformity
- Flexed, abducted, externally rotated hips, often with dislocation
- Flexion deformity of the knee
- Club foot deformity

About the Disorder

Arthrogryposis is a musculoskeletal disorder characterized by the presence of multiple joint contractures (limitation of the range of motion of a joint) at birth. In some cases, only a few joints may be affected. However, in the classic cases of Arthrogryposis Multiplex Congenita (AMC), hands, wrists, elbows, shoulders, hips, feet, and knees are affected. In the more severe cases, joints in the back and jaw can be affected as well. In addition to having joint contractures, children also experience muscle weakness, which further limits movement.

AMC is a nonprogressive congenital neuromuscular syndrome characterized by severe joint contractures, muscle weakness, and fibrosis. AMC occurs in 1 out of every 3,000 live births. There may be as many as 10 to 20 different arthrogryptic disorders, all with similar joint manifestations. The most common form (43%) of AMC is amyoplasia, characterized by fatty and fibrous tissue replacement of the limb muscles.

There are many different causes of AMC, but typically it is a result of either problems with joint growth and development, decreased fetal movement (not enough room in the uterus to move), or problems with spinal development in the first 3 months of pregnancy. A diagnosis of AMC can sometimes be made during pregnancy. Ultrasounds at approximately 20 weeks gestation may show abnormal position of joints or lack of movements in joints and limbs indicating the disorder. Otherwise, the diagnosis can be made by an orthopedist based on clinical symptoms and findings. Muscle biopsies, blood tests, and clinical findings help rule out other possible disorders and provide evidence for AMC.

The primary joints involved (in order of decreasing prevalence) include the foot, hip, wrist, knee, elbow, and shoulder. AMC is typically symmetrical and involves all four extremities with some variation seen.

Typically, two forms are seen. First form: Flexed and dislocated hips, clubfeet (talipes equinovarus), extended knees, flexed elbows, flexed wrists and fingers. Second Form: abducted and externally rotated hips, flexed knees, clubfeet, internally rotated shoulders, extended elbow, pronated forearm, and flexed and ulnarly deviated wrists.

Other associated conditions include scoliosis, lung hypoplasia (underdevelopment) leading to respiratory problems, growth retardation, midfacial hemangioma (benign tumor of dilated blood vessels), facial and jaw deformities, respiratory problems, and abdominal hernias. Cognition and speech are usually normal.

There is no cure for AMC; however, there are treatments that can help children live very full lives. For many, physical therapy has proven to be beneficial to strengthen muscles and improve range of motion. Splinting and bracing can also help improve range of motion. If these traditional treatments have not produced positive results, surgery may be necessary. Surgery can be done to put feet in position for standing and walking and can also be done to knees, hips, elbows, and wrists to achieve greater range of motion. And, in some cases, muscle and tendon transfers can be done to improve range of motion. AMC is not a progressive disorder, so it will not worsen with age. However, children must receive medical treatment to prevent joints from stiffening as they grow. Most children with AMC have some walking ability. Bracing and other treatments mentioned above will help to make walking easier. AMC may be accompanied by other disorders, such as central nervous system disorder. However, in most cases, the long term outlook is positive. Most individuals with AMC possess normal intelligence and speech. They have a potential for functional mobility and are able to lead productive, independent lives.

Educational Implications

Children with AMC usually possess normal intelligence and speech. The student may need support in dealing with self image and acceptance from his/her peers. Physical therapy, occupational therapy, and adaptive physical education will be needed to address the student's specific needs. Assistive equipment may be necessary as well as an adapted classroom environment. Due to limited mobility, the student may need additional hall passing time. The student may need academic services provided when absent for long periods of time due to scheduled surgeries.

Instructional Strategies and Classroom Accommodations

Staff may need to provide:

- Curriculum modifications (extra time for assignments, modifications to length of assignment, no time limits, alternative ways to test and get information, adapted materials and environment, extended passing time, support from resource room) and possible support from PT/OT/DAPE.
- May need assistance at lunch.
- May not be able to write or inability to write as fast as other students. The student may need to do work orally, use a computer for written assignments, may need alternative note taking methods, or other handwriting accommodations.
- May become tired easily from walking or other motor activities. Should be allowed more time to get from one place to another.
- May need assistance with daily classroom activities such as getting books out of book bag, etc.
- Check to see if the student can walk up steps.
- An Individual Health Care Plan and Emergency Evacuation Plan should be developed and implemented.

Resources

Avenues

<http://www.avenuesforamc.com>

Kidshealth, The Nemours Foundation

<http://www.kidshealth.org/faq/arthrogryposis.html>

E-mail: NIAMSBoneInfo@mail.nih.gov

Wheless' Textbook of Orthopedics

<http://www.medmedia.com/012/110.htm>

Orthoseek

<http://www.orthoseek.com/articles/arthrogryposis.html>

The Medline Database

The National Rehabilitation Information Center

National Organization for Rare Diseases

NIH/National Arthritis and Musculoskeletal and Skin Diseases Info. Clearinghouse

<http://www.niams.hih.gov>