

Arthrogryposis Multiplex Congenita: A Clinical Image Report

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Arthrogryposis multiplex congenita (AGC) is a heterogeneous group of non-progressive conditions characterized by multiple joint contractures [1, 2] which may involve the body at birth [3, 4]. It is known to be typically symmetrical and is also known as amyoplasia [5]. It presents either in isolation or in association with other syndromes. Its etiology is considered multifactorial but poorly understood [6]. Its occurrence has shown no racial or sexual predisposition, except sporadic cases that were seen commonly in isolated populations in Finland and the Bedouin community in Israel [7]. Implicated etiological factors include chromosomal abnormalities, maternal viral infections such as enterovirus and roseola virus, and attempted termination of pregnancy, as reported by Wynne and Roberts [8]. It is usually diagnosed with muscle biopsy [9] and electromyography. Management of AMC is multidisciplinary [9]. The images of the indexed

Figure 1: Right wrist

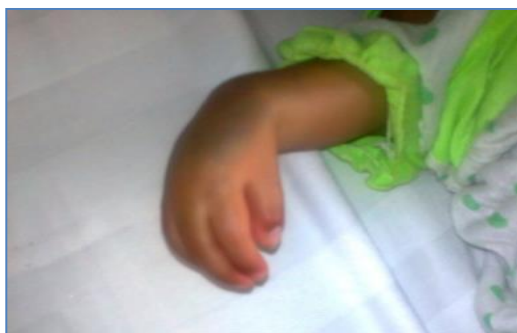


Figure 2: Anterior view of right hip

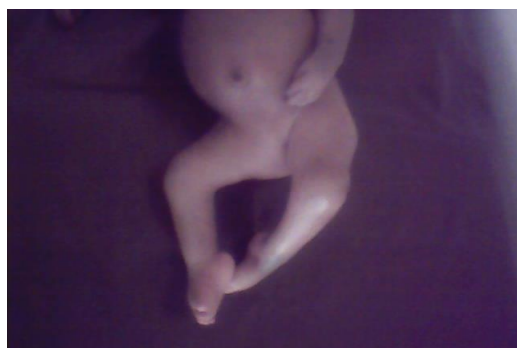
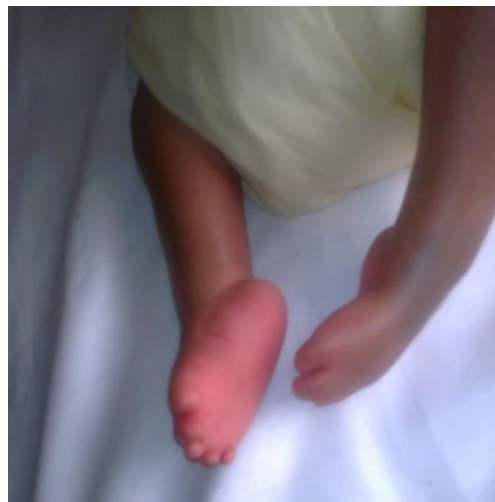


Figure 3: Congenital talipes equinovarus of both hips



case were used to demonstrate some clinical features of AGC as well as the atypical hip asymmetry.

A month-old female child was brought to the orthopedic clinic by her 20-year old mother, who noticed global hypertonia, hip-hip bulk discrepancy, and abduction-flexion deformity of the right hip since birth. The mother had attempted termination of pregnancy at 9 and 12 weeks of gestation by use of unknown oral and injectable medications. Patient's mother received no antenatal care and birth was supervised by a traditional birth attendant. Infant had been immunized with oral polio vaccine and BCG vaccine, and was breastfed at the time of presentation with water supplementation.

There was a discrete, non-discharging erythematous, non-itching, papular skin rash and right upper limb showed marked hypertonia with fixed flexion deformity of the hand at the wrist (Figure 1), while left side showed marked hypertonia of the arm with fracture of the humerus (Figure 4), and no active movement at the shoulder region.

The left lower limb had marked hypertonia with no active movement at the hip and knee while right lower limb had excessive range of movem-

ent at the hip. There was hip-hip discrepancy in muscle mass (Figure 2), abduction/flexion deformity of the right hip, plantar-flexion with inversion deformity of both feet (Figure 3) and bilateral brachydactyly. Spine was intact with marked hypertonia in opisthotonic position and presence of good sucking reflex. There was absence of labia in the perineum and iris was dark brown. X-ray film showed united fracture in mid-shaft of left humerus (Figure 4), and angular deformity with apparent subluxation of adjacent elbow joint (Figure 4). Figure 5 shows hip-hip disproportion and joint asymmetry. A diagnosis of arthrogryposis multiple congenita was made, and the mother was counseled concerning the condition of her baby. Intensive physiotherapy was commenced. Follow-up visit at 3 weeks showed adequate weight gain of 1.0 kilogram. Head circumference was noted to be 37 cm, which was adequate for age.

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Figure 5: Anteroposterior view of abdomenopelvic region

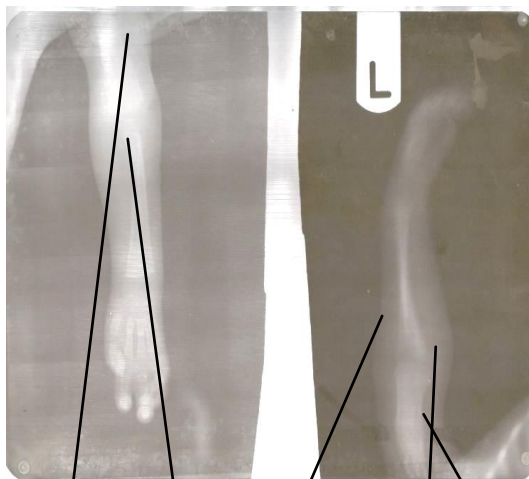


Axis of ossified bones

Left hip appears normal with close proximity of areas of ossification

Right hip dislocation with far displacement of areas of ossification

Figure 4: Anteroposterior and lateral view of left upper limb



Old united fracture

Soft tissue swelling

Subluxation of elbow joint

Old united fracture

Subluxation of joints/angular deformity