

Radiographic changes in arthrogrypotic knees

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Abstract. We evaluated the knees of 62 patients with arthrogryposis multiplex congenita radiographically. Abnormal radiographs were noted in 34. The abnormalities consisted of both congenital and chronic changes secondary to long-standing clinical deformities including patellar elongation, malposition, flattening of the femoral condyles, joint incongruity, tibial plateau irregularities, tibial and femoral fractures, fibula hypoplasia, soft-tissue thickening, valgus deformity, and dislocation. The radiographic findings were consistent with the degree of longstanding physical deformity and can be used as a guide to the severity of the condition and the need for treatment.

Key words: Arthrogryposis – Knee – Chronic changes – Malposition

Arthrogryposis multiplex congenita is a noninherited syndrome of multiple fixed contractures occurring at birth, with an incidence of 1 per 3000 live births. The term is derived from the Greek meaning “curved joint,” and the entity was originally described by Otto in 1841. There is a 1:1 male to female ratio [1]. Arthrogryposis multiplex congenita most probably represents a spectrum of diseases rather than a single entity.

Numerous etiologies have been proposed including mechanical factors, neurologic lesions, and muscle abnormalities [2, 3, 6]. Despite disagreement over the precise mechanism leading to this syndrome, it seems clear that decreased fetal movement, a common outcome of all these etiologies, contributes to the multiple joint contractures that are pathognomonic of arthrogryposis multiplex congenita.

The arthrogrypotic child presents with a typical clinical picture [2, 8]. Sensation and intelligence are normal, but muscle mass is decreased and the extremities are long and slender. There are diminished skin creases and subcutaneous tissue with webbing or pterygium posterior to the joints. The periarticular tissues and joint capsules are contracted and fibrotic, contributing to the de-

formity. The knees present with flexion contractures or, less commonly, extension deformities. Dislocations or malposition of the knees is also common [7].

Treatment of these patients consists of casting, bracing, physical therapy, and surgery to relieve contractures. The deformities are frequently severe and recurrent and lead to long-standing clinical and radiographic abnormalities.

Previous studies have reviewed the clinical findings of these patients, but few have concentrated on the knee abnormalities or the associated radiographic findings [5, 7]. This paper specifically deals with the radiographic manifestations of arthrogrypotic knees.

Materials and methods

A total of 46 patients with arthrogryposis multiplex congenita were reviewed at the Shriners Hospital for Crippled Children, Tampa Unit. Forty suffered knee joint involvement, and 22 had demonstrable radiographic abnormalities consistent with chronic or con-

Table 1. Radiographic knee abnormalities in 34 patients with arthrogryposis multiplex congenita

Abnormality	No. of knees
Elongated patella	15
Absent patella	6
Hypoplastic patella	4
Patellar dislocation	1
Patella alta	3
Patella baja	4
Patellar fracture (proximal pole)	1
Patellar fracture (distal pole)	3
Triangular patella	1
Flattened femoral condyle	9
Atrophic femoral condyle	1
Irregular femoral condyle	2
Femur fracture	3
Tibial fracture	9
Dome-shaped tibial epiphysis	5
Flat/irregular tibial plateau	8
Fibula hypoplasia	4
Hyperplastic proximal fibula	1
Knee dislocation	1
Recurvatum	1
Valgus deformity	1
Periarticular thickening	13
Popliteal webbing	1

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Fig. 1. Patella elongation secondary to flexion contracture occurred in 15 patients with both upper and lower pole fractures. These are similar to patella sleeve fractures, with the periosteum and ligamentous attachments pulling off the patella pole

Fig. 2. In one patient a triangular patella was noted secondary to contracted soft-tissue forces exerted longitudinally, medially, and laterally

Fig. 3A, B. Late degenerative changes included femoral condylar irregularity, dome-shaped tibia and hypoplastic fibula, and patella elongation with baja position

Fig. 4A, B. Development of degenerative changes from infancy (A) to age 12 years (B) is demonstrated in this patient. There is

persistent rotation, soft-tissue contracture, and absence of the patella. There are thickened soft-tissue densities anteriorly but no evidence of patella ossification by age 12

Fig. 5. Knee dislocation in a newborn, similar to Larsen's syndrome but lacking the constellation of skeletal dysplasia, facial changes, and multiple dislocations

Fig. 6. Fractures occurred frequently in arthrogryptic patients due to osteoporosis, severe soft-tissue contracture, and manipulation of the extremities. Several fractures in the younger patients were not diagnosed initially, and exuberant callus formation resulted as shown here

genital arthrogryptic deformity. We examined all patients clinically and radiographically, and the knees were grouped according to the radiographic findings. In addition, 16 radiographs of previous patients with arthrogryposis multiplex congenita from Newington Children's Hospital, Newington, Connecticut, were considered. Of this group, twelve showed knee abnormalities, yielding a total of 34 patients for radiographic review.

Arthrogryposis multiplex congenita probably represents a spectrum of disease states defined by the presence of multiple, non-progressive joint contractures at birth. The patients presented with initial diagnoses including: arthrogryposis multiplex congenita, pterygium syndrome, amyoplasia congenita, chromosomal abnormalities, and caudal regression syndrome. The 34 patients ranged in age from 2 months to 21 years, with a mean of 7.2 years.

Results

Radiographically patellar abnormalities were the most common and were noted in 29 of 34 patients (Table 1). Patellar elongation was seen in 15 patients with long-standing flexion contractures. In these radiographs the patella appeared stretched over the anterior aspect of the deformed knee joint (Fig. 1). In 4 patients the patella was fractured at its proximal (1 patient) or distal (3 patients) pole. The patellar pole fracture is similar to the patellar sleeve fracture described by Ogden [4]. An abnormal position of the patella was seen in 3 knees with patella alta and in 4 with patella baja. The patella and/or its ossification center were completely absent in 6 patients, hypoplastic in 4, and triangular in 1 (Fig. 2).

Femoral abnormalities included condylar flattening in 9 patients, atrophy from poor contact in 1, and articular surface irregularities in 2 (Fig. 3). Tibial changes were also noted, with 5 knees demonstrating a dome-shaped proximal tibial epiphysis. Flattening of the tibial plateau was identified in 7 patients, and another exhibited marked articular surface irregularities (Fig. 4). Associated fibular anomalies included hypoplasia in 4 patients and hyperplasia in 1.

Increased soft-tissue density was noted radiographically in 13 knees, reflecting the thickened nature of the joint capsules. This manifested as thickened soft-tissue planes with distinct margins. One patient each showed recurvatum, valgus deformity, knee dislocation (Fig. 5), and popliteal webbing.

Fractures occurred in 3 femurs and 9 tibias. Two of the tibial fractures involved the plateau area. All were treated conservatively, and some presented with associated osteoporosis after casting or surgery. In newborns, several fractures in both the tibia and femur were diagnosed after manipulation and casting of contractures (Fig. 6).

Discussion

Thomas et al. and Williams have previously evaluated knee changes in patients with arthrogryposis [7, 8]. Both studies noted knee flexion and extension contractures, ligamentous instability, and recurvatum. In the present series knee radiographs of 62 patients with arthrogryposis multiplex congenita were evaluated, 34 of whom were seen to have radiographic abnormalities. The changes observed in the joints were found more commonly in those patients with greater magnitudes of contracture and correlated with the longstanding nature of the deformity and the age of the patients.

The patella was the most commonly involved bone of the knee. Elongation was seen in 15 patients. The crouched gait with knee flexion seen in many arthrogryptic children exerts abnormal tension on the patella, which leads to elongation. The same mechanism is also probably responsible for fractures of the patella pole [4].

A triangular patella (Fig. 2) was noted in one patient in this study. This appearance probably represents a

combination of longitudinal elongation and transverse broadening as a result of excessive tightness of the retinacular tissues. Subluxation of the patella was a rare finding in this patient population due to the contracture of the capsule. Patellar ossification, which normally begins before age 4 was significantly delayed in 4 patients. This delay produced the appearance of a hypoplastic patella. The patella was totally absent in an additional 6 patients (Fig. 4), perhaps secondary to the lack of developmental stimulus from an incongruous and stiff joint.

The femoral condyles were irregular in contour in 12 patients. Fractures of the distal femur may have been responsible in 2 patients. Another 9 showed evidence of flattening of the femoral condyles, probably due to the abnormal apposition of the bony articulations. The increased pressures exerted by the contracted knee may lead to the observed compensatory remodeling of the distal femur (Fig. 3).

In 5 patients with long-standing contractures, molding of the proximal tibia produced a dome-shaped epiphysis (Fig. 3). This appears to be a result of decreased contact, motion, and pressure in these deformed knees. Alternatively, 7 tibial plateaus exhibited flattening, presumably from long-term increased pressure.

Fractures can occur in these patients secondary to osteoporosis, soft-tissue contracture, and manipulation of stiff joints (Fig. 6). The fractures may be missed in the newborn population, and marked callus formation may result. Fortunately, remodeling occurs readily in these patients.

Radiographic evaluation demonstrates the marked molding effect secondary to abnormal placement or pressure on the femur, tibia, and patella in the arthrogryptic knee. This analysis aids evaluating intraarticular pathology and its duration and may serve as a guide to treatment or surgical intervention. Based on this review; we feel that early surgical treatment of the arthrogryptic knee deformity is indicated to prevent chronic changes. Knowledge of these types of abnormalities is essential for the orthopedic surgeon and the radiologist in planning the care of these complex patients.

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