Muscle ultrasonography in arthrogryposis

Comparison with clinical, neuromyographic and histologic findings in 41 cases

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We studied the ultrasonographic appearance of muscles in 41 patients with arthrogryposis multiplex congenita (AMC). The observations were compared to the findings in clinical testing, electroneuromyography, and histological examination. The strength of the muscles in AMC-patients did not always correspond to the ultrasonographic morphology of the muscles. In extension contractures of the elbow and in equinovarus adductus deformities of the foot there was a distribution of pathological changes in the muscles corresponding to the primary deformity. The sound transfer through the muscles was decreased, notably in the neurogenic form of AMC.

Ultrasonography is an important complement to the clinical examination and it should always be done prior to surgery and muscle biopsy.

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Arthrogryposis multiplex congenita (AMC), a congenital disorder characterized by multiple joint contractures, which according to Wynne-Davies (1981) can be divided into a neurogenic and a rare myogenic form. Abnormal muscles are often found (Gibson et al. 1970) and when causing muscular imbalance, it will probably be important in determining the type of deformity, flexion and extension contractures, respectively (Brown et al. 1980). Difficulties are often encountered in the evaluation of the muscular status of these patients. A needle biopsy from the muscles of a patient with neurogenic arthrogryposis may show normal muscular tissue, fibrous replacement and morphological changes characteristic of chronic denervation, depending on the site from which the biopsy is taken (Hageman et al. 1983).

Electroneuromyography (ENMG) can be used for detection of atrophy of the alfa-motorneurons (Amick et al. 1967). This method has its limitations; totally atrophied muscles, from which no electrical potentials can be recorded, can be scattered with obviously normal muscles, which has normal innervation. Ultrasonography of muscular tissue has many clinical applications (Heckmatt et al. 1982, Young and Hughes 1982, Risseeuw et al. 1985, Lamminen et al. 1988). It has been used among others in morphologic examination of patients with spinal atrophy, a condition, that with respect to the muscular changes, resembles AMC (Lipparini et al. 1985). Our purpose was to test the usefulness of ultrasonography in the assessment of muscular status in AMC.

Patients and methods

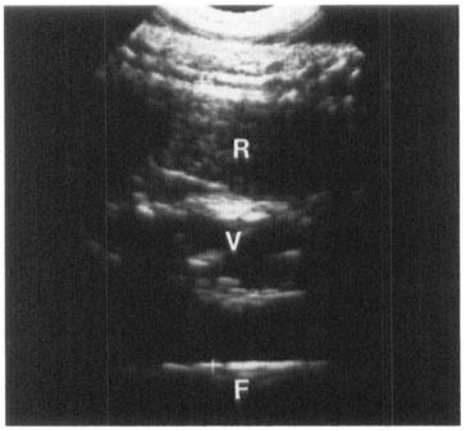
In our hospital, 62 patients with AMC were treated during 1949–86. In connection with a follow-up examination, 41 patients had ultrasonography of selected muscles (Table 1). 11 patients had a neurogenic form of AMC, 1 patient had a myogenic form, and in the rest of the patients no specific type could be identified.

A sector ultrasound apparatus (ATL 100) with 7.5 MHz Scanhead was used, and the images were recorded on monitor film. The transducer was placed on a 1-cm-thick gel pad (Kiteko) to get a noncompressing contact with the skin. Transmission gel was

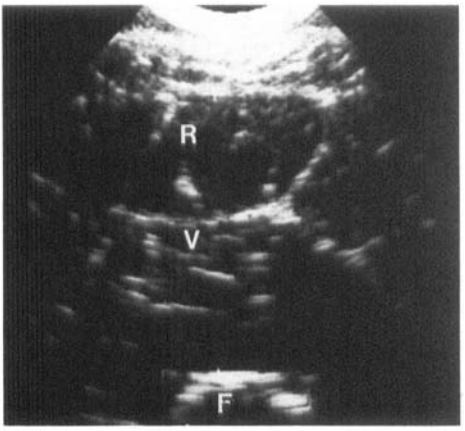
Table 1. 41 AMC-patients examined by ultrasonography

Involved extremity	Men	Women	Age range
Upper	2	3	0.3–12
Lower	2	8	15–28
Both	18	8	0.1–33

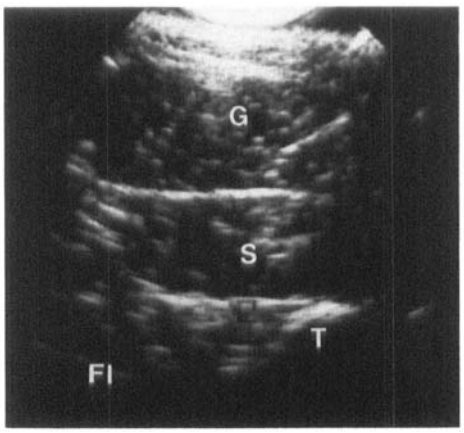
Figure 1. Ultrasonography of normal muscles. F femur, T tibia, Fl fibula.



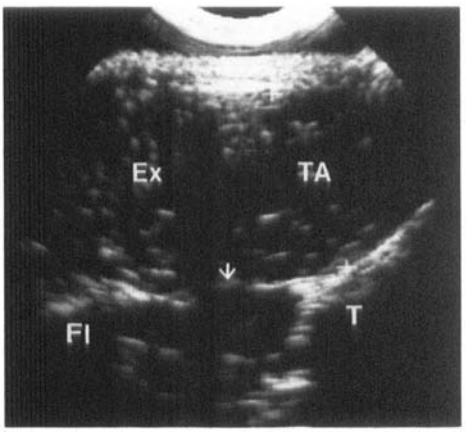
A longitudinal section of normal rectus femoris (R) and vastus intermedius muscles (V).



A cross-section of normal rectus femoris (R) and vastus intermedius muscles (V).



A cross-section through the posterior calf. Gastrocnemius (G) and soleus (S) are normal.



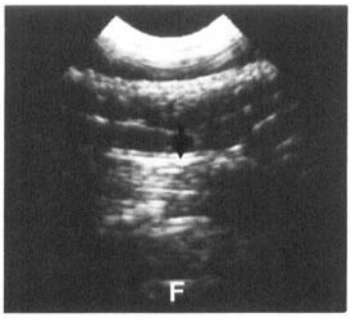
A cross-section through the anterior compartment of the leg. Normal structures of the tibialis anterior (TA) and extensor digitorum muscles (Ex) are evident and the intermuscular septa and the membrana interossea (arrow) can also be detected.

used for acoustic coupling. Transverse and longitudinal scans were obtained from anterior and posterior aspects of the midthigh and midcalf, respectively, and anterior and posterior aspects of the midlevel humerus. The ultrasonographic findings were graded into diffuse and local distributions. The ultrasonographic appearance of each muscle was recorded and graded 0–3: 0) normal appearance; 1) sound transmission slightly disturbed. Muscle fiber can be identified. Intermuscular septal structure normal; 2) sound transmission clearly disturbed, showing increased echo density. Fiber structure abnormal. Septal structure evident; 3) sound transmission severely disturbed, displaying decreased depth of penetration. Fiber structure not visible, intermuscular septal structure indistinguishable. Muscle mass diminshed (Figures 1–3).

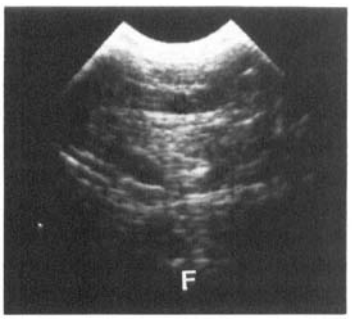
The primary joint status was recorded from the hospital charts. At the follow-up the patients were examined clinically. The mobility of the joints was coded and the muscle strength was tested and graded 0–5 according to Daniels and Wortingham (1972).

In the clinical examination of the patients, special attention was paid to testing the muscles acting on the knees and elbows. The clinical testing of these muscles is relatively easy and they are well visualized with ultrasonography. The clinical testing showed that there were 10 knees which had no functioning extensor muscles and there were 13 elbows in which the extension function was lacking. The ultrasonographic examination did not include all muscle groups in all patients; the quadriceps muscle was investigated in all patients, the biceps femoris muscle in 38 legs. 15

Figure 2. Ultrasonographic findings in a patient with flexion contractures of the knees. There is diffuse increased echo density and the change is Grade 2.

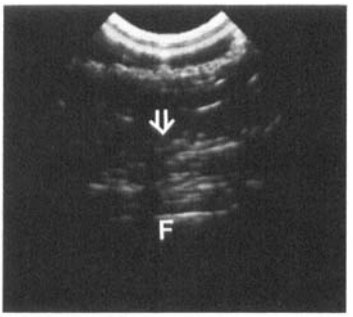


A longitudinal scan at the midlevel of the right femur. Rectus femoris and vastus intermedius are seen between femur (F) and the arrow.

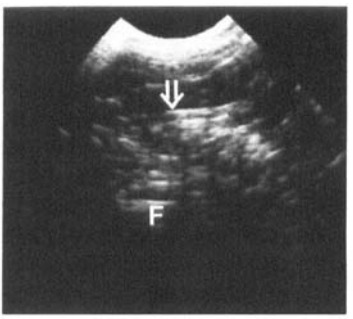


A transverse scan at the midlevel of the right femur.

Figure 3. Ultrasonographic findings in a patient with flexion contractures of the knees. There is increased echo density, intermuscular septa are indistinguishable and muscle mass is diminished. The changes are severe, Grade 3.

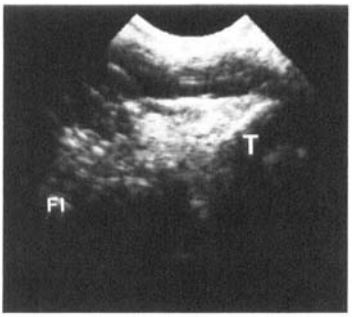


A longitudinal scan.The muscle mass of rectus femoris and vastus intermedius is seen between the femur (F) and the arrow.

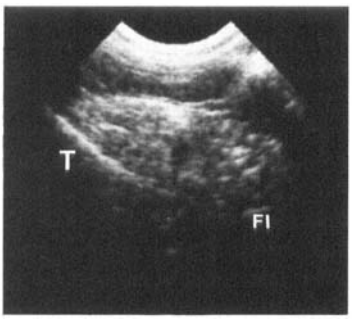


A transverse scan.

Figure 4 Ultrasonographic findings in the anterior compartments of the leg, showing diminished muscle mass and increased echo density of the right side (Grade 3) and less involvement of the left side (Grade 2).



A transverse scan of the right anterior compartments of the leg.
T tibia, Fl fibula.



A transverse scan of the left anterior compartments of the leg.

arms, usually in cases with normal findings in the contralateral arm, were not examined with ultrasonography. Electroneuromyographic and histologic findings were recorded. The results of the ultrasonographic examination were compared with clinical muscular status and with results of other examination methods.

There were 3 patients with almost normal ultrasonography in all muscle groups (Figure 1), the only changes being a Grade 1 finding of either the peroneal or gastrocnemius muscles. Grade 3 pathology was seen in altogether 30 muscles, diffuse in 27 cases and local in 14.

A normal muscle power was not always accompanied by a normal ultrasonography (Table 2). Indeed, there were 10 clinically normal muscles, for which the ultrasonographic findings were Grade 2. On the other hand, there were 11 muscles, clinically tested as nonfunctioning, with an ultrasonographic Grade 1.

The different types of primary deformities were compared to the ultrasonographic findings (Table 3). A distribution of the muscular pathology mechanically

Table 2. Correlation between the clinical testing and ultrasonographic findings in extensors and flexors of 67 elbows and in 82 extensors and 38 flexors of the knees

Muscular power (0–V)	Ultrasonographic grade 0–3			
	0	1	2	3
V	89	25	10	0
IV	3	17	5	6
III	0	9	8	1
II	0	5	9	6
I	0	9	8	8
0	1	11	15	9
Total	93	76	55	30

corresponding to the primary deformity was seen only in the elbow extension contracture group, which had a higher degree of pathology in the flexors of the elbow. The group with pes equinovarus had generally a more severe involvement of the muscles (Table 4); the mus-

Table 3. Relationship between the primary deformity and the ultrasonographic findings in the extensor and flexor muscles of the knees and elbows. Only patients whose examination included both extensors and flexors are presented. Figures show no. of extensor/no. of flexors

	Ultrasonographic Grade				
Contracture	0	1	2	3	mean
Knee extension (9 pts, 18 knees)	2/4	12/12	4/2	0/0	1.1/0.9
Knee flexion (5 pts, 10 knees)	0/0	3/4	4/3	13/3	2.0/1.9
Elbow extension (13 pts, 23 elbows)	5/2	8/6	10/9	0/6	1.2/1.8
Elbow flexion (9 pts, 17 elbows)	2/1	2/2	5/8	8/6	2.1/2.1

Table 4. Ultrasonographic appearance of the muscles of the anterior compartment of the leg and the gastrocnemius muscles in 61 feet with planovalgus and equinovarus adductus deformities

	Planovalgus feet		Equinovarus	
Grade	Ant comp	Gastr	Ant comp	Gastr
0	6	7	4	6
1	5	4	26	27
2	0	0	19	17
3	0	0	1	0
Total	11	11	50	50

Table 5. Ultrasonographic changes and their correlation to the histopathologic findings in the muscles

		Ultrasonographic Grade			
Histology	n	0	1	2	3
Neurogenic atrophy	2	1	1		
Congen fiber disproportion	1	1			
Unspecific	2	1			1
Normal	12	6	2	3	1
Total	17	9	3	3	2

cles of the anterior compartment (tibialis anterior muscle and long extensors of the toes) having a slightly higher degree of pathology (Figure 4). 12 patients had a lower motoneuron affection demonstrated in ENMG. Of these, 1 had diffusely distributed ultrasonography changes. 17 muscle biopsies were taken, all without guiding by ultrasonography. Out of 12 patients with normal histology only 6 had normal ultrasonography of the biopsied muscle (Table 5).

Discussion

The patients in this study represent an unselected material of AMC-patients. The etiological background to the syndrome in this study was in accordance with the results of Wynne-Davies et al. (1981).

The clinical testing of the muscles in AMC-patients is difficult for several reasons, e.g., joint contractures, use of compensatory muscles, and multiply-involved muscle groups. The subcutaneous tissue is often thick and makes the extremity look cylindrical. Muscular atrophy is therefore easily overlooked and active muscle contractions can be difficult to observe. The difficulty in clinical testing of muscles in these patients is clearly demonstrated in the high number of structurally abnormal muscles with good function. On the other hand, there were also a substantial number of clinically poor muscles with normal or nearly normal structure.

Information about the muscular status is needed, especially when a muscle transfer is planned, as in the cases of restoration of elbow flexion (Holtman et al. 1975). In our material there were 43 elbows with a clinically well-functioning triceps brachii muscle (strength 4–5). Of these, 6 muscles had an increased echo corresponding to an alteration in the muscle structure of Grade 2 or 3. These muscles would probably not be suitable for a transfer. On the other hand, there were 12 elbows with no demonstrable function in the triceps, and in 4 of these there were few, if any, structural changes on ultrasonography. In these cases there was a rigid flexion contracture of the elbow.

According to Brown et al. (1980) there should be more structural changes in flexor muscles of patients with extension contractures, and in patients with flexion contractures there should be more structural changes in extensor muscles. This was, however, not true in knee deformities, but in extension contractures of the elbows the majority of the patients had more pathological changes in the flexor muscles. It is possible that the type of deformity is determined also by other factors than muscular imbalance.

The contribution of the ultrasonography to the etiological typing of AMC is rather complementary to other methods. In the typical form of neurogenic AMC the muscle changes were diffusely distributed, more often of Grade 2 or 3, and often an increase in the depth of the subcutaneous fat tissue was observed. The muscular atrophy was also more marked in these patients.

Varying degrees of muscular involvement demand precise planning of the site of the biopsy. A surgical nonselective biopsy may show a normal histological structure of a muscle which is partly abnormal. In this study there were 12 normal histological findings; in 11 of these cases ultrasonographic guidance would have

contributed to more abnormal tissue. The limitations of the use of ultrasound in these patients are few. It gives, especially in a young child, a good survey of the severity of the condition. Ultrasonography is easily performed and gives additional information about distribution of the muscular involvement, which helps to plan diagnostic and surgical procedures.

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