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Real-Time Ultrasonography in Neuromuscular Problems of Children

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Abstract: Ultrasound imaging of 20 cases of progressive muscular dystrophy and 10 cases of suspected infantile spinal muscular atrophy in children was performed by us, as a double-blind pilot study matched against 25 controls. Open muscle biopsy was restricted to the muscular dystrophy group. The ultrasonographic findings were correlated with parameters such as functional disability of muscle and muscle biopsy features in the dystrophy group. It was interesting to observe that the muscle echogram was abnormal in both types of neuromuscular problems, the controls giving a normal muscle echogram. Ultrasonography was helpful in detection of unequivocal changes in our cases with mild clinical disability. It had a close correlation with changes in gross muscle architecture, as seen on muscle biopsy. **Indexing Words:** Ultrasound · Neuromuscular problems

Real-time ultrasonography is a simple, noninvasive procedure that is most suitable for application in pediatric practice. The usefulness of real-time ultrasonography in assessment of neuromuscular problems in children has gained importance only in recent years, and there is thus very little literature on it.[1] The ultrasonographic appearance of various disorders in children such as progressive muscular dystrophies, infantile spinal muscular atrophy, congenital myopathies, and motor neuropathies has been found to be strikingly abnormal.[2] Interpretation of ultrasonic scan findings depends on the echogenicity of muscle compared with the echogenicity of bone, as matched with controls. The normal ultrasonic scan of muscle shows a striking bone echogenicity in contrast to a relatively echo-free muscle zone.

In this article, we have done a pilot study using real-time ultrasonography in children with progressive muscular dystrophy and in infants with a clinical diagnosis of infantile spinal muscular atrophy in an attempt to correlate their clinicopathologic profiles with scan findings.

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MATERIALS AND METHODS

Twenty children with progressive muscular dystrophy, including 19 boys and one girl, and 10 infants with hypotonia and areflexia from birth who were seen at the Paediatric Neurology Outpatient Department of the Institute of Child Health and Hospital for Children, Madras, were examined by ultrasonography. Fifteen normal children in the age group of 4–15 years and 10 infants in the age range of 2 months to 1 year were studied as matched controls. In the dystrophy group, 17 children had an onset of illness after the age of 5 years, two cases before 10 years, and only one case before 2 years. All were above the age of 5 years. In one family, three boys were affected, two of whom were examined. The infants presenting with areflexic hypotonia and delay in motor milestones ranged from 3 months to 1.5 years of age at the time of scan.

A thorough general examination with emphasis on the spinomotor system was made in each case. In cases with muscular dystrophy, serum creatine kinase estimation, chest x-ray, electrocardiogram, electromyogram, and open muscle biopsy from quadriceps femoris were done.[3,4] The muscle biopsy findings were accorded a score as follows: grade I, normal architecture of muscle with no infiltration of fat or connective tissue; grade II, some invasion of fat or connective tissue; grade III, disruption of muscle fascicles; and

grade IV, severe changes with >50% infiltration by fat and connective tissue.[2] Intelligence quotient assessment was done only in children with clinical submentation. In infants with clinical suspicion of Werdnig Hoffman's disease, serum creatine kinase measurements, and electromyograms were done. Muscle biopsy was not done in these cases. Both groups of children along with normal children were subjected to ultrasonographic study of muscles.

A real-time B-mode ultrasound scanner (ALOKA-SSD 256) was used. The frequency of the transducer was 5 MHz, with varying standardization according to age. Scans were made over the thigh in both longitudinal and transverse directions. Scanning was done using a water bath at room temperature to help in delineating the curvature of thigh, which is otherwise not possible with a rigid transducer, and to give clearance between the transducer and the site of scanning. The intensity of the echoes reflected from muscle was classified on a four-point scale. Grade I is normal scan, grade II a scan in which there is an increased muscle echogenicity with distinct bone echogenicity, grade III a scan in which there is a marked increase in muscle echogenicity and reduced bone echogenicity, and grade IV a scan in which there is very strong muscle echogenicity with complete loss of bone echogenicity.[2]

RESULTS

Normals

The ultrasonic scan findings in normal children gave similar results in that all showed a strong bone echo with fairly anechoic muscles. The boundaries between the vastus intermedius and vastus lateralis as well as the fascia lata and subcutaneous space were clearly seen (Figures 1 and 2).

Progressive Muscular Dystrophy

In the progressive muscular dystrophy group, the duration of disability was from 4 months to 6 years at the time of the ultrasound examination. Fourteen children had involvement of both pelvic and pectoral girdle muscles and the rest had weakness of the pelvic muscle group. Except for three cases, all had varying degrees of muscle wasting. There was no muscle tenderness or fasciculation in any patient. Eighteen cases had marked pseudohypertrophy of the calves. Muscle power varied from grade 2 to 4 on the MRC scale (Table 1). Deep tendon reflexes were sluggish to absent with preservation of ankle jerks. The intelligence quotient was ≤80 in six children. The

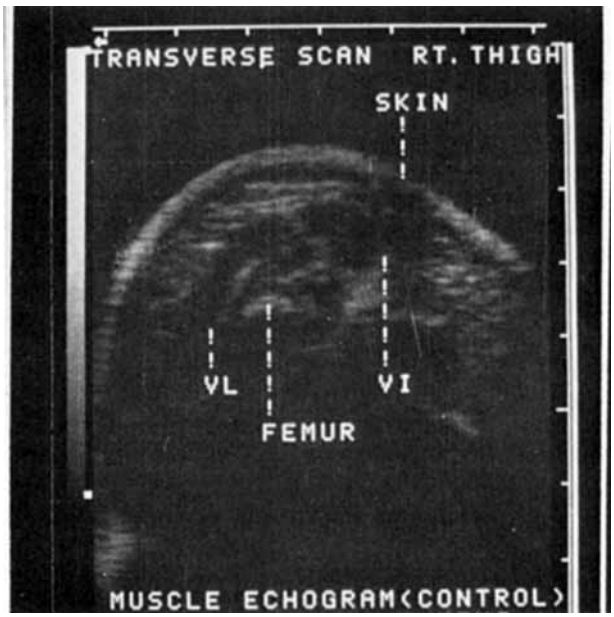

FIGURE 1. Transverse scan of thigh in a 2-month-old normal infant showing distinct bone echo, relatively echo-free muscle groups, and normal subcutaneous space. VL, vastus lateralis; VI, vastus intermedius.

serum creatine kinase levels were high in all cases. Five children had electrocardiographic abnormalities. The electromyogram showed normal insertion activity with myogenic pattern in all of these cases. The muscle biopsy showed varying stages of muscle degeneration from the early stages of variation in muscle fiber size, rounding of fibers, and intact cross striation to gross replacement by fibrofatty tissue with no semblance of muscle architecture.[3-5]

The ultrasonic scan was abnormal in all cases

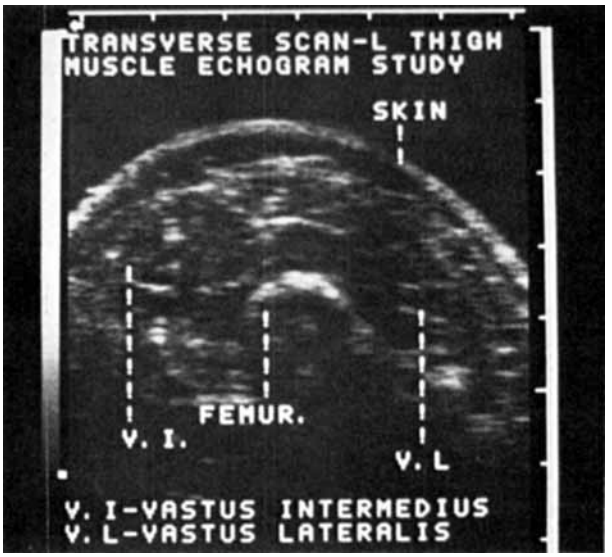

FIGURE 2. Transverse scan of thigh in a 4.5-year-old normal boy showing distinct bone echo, relatively echo-free muscle groups, and normal subcutaneous space.

TABLE 1
Correlation of Clinicopathologic Findings with Muscle Echogenicity Score in Cases of Progressive Muscular Dystrophy

Case no.	Functional MRC grading	Muscle biopsy score	Muscle echogenicity score	Delineation of muscle groups
1	4	II	II	+
2	3	IV	IV	–
3	4	II	III	+
4	3	II	II	+
5	3	III	III	–
6	2	IV	III	+
7	4	II	II	+
8	2	IV	IV	–
9	2	IV	IV	–
10	4	III	IV	–
11	4	IV	III	–
12	2	IV	IV	–
13	2	III	III	–
14	4	III	III	–
15	3	IV	III	–
16	3	IV	IV	–
17	2	IV	IV	–
18	3	II	II	+
19	1	IV	IV	–
20	3	III	IV	–

of progressive muscular dystrophy (Table 1). It showed grade III–IV changes in 16 cases and grade II changes in four cases (Figures 3–5). The delineation of muscle groups was less obvious in cases beyond grade III (Table 1). There was a relationship of the ultrasound image to the severity of pathologic changes in these children. The end-stage findings in muscular dystrophy correlated with grade IV scans (Table 1). The early-stage findings corresponded to a grade II scan. There was, however, no correlation of scan findings with functional disability in our cases (Table 1).

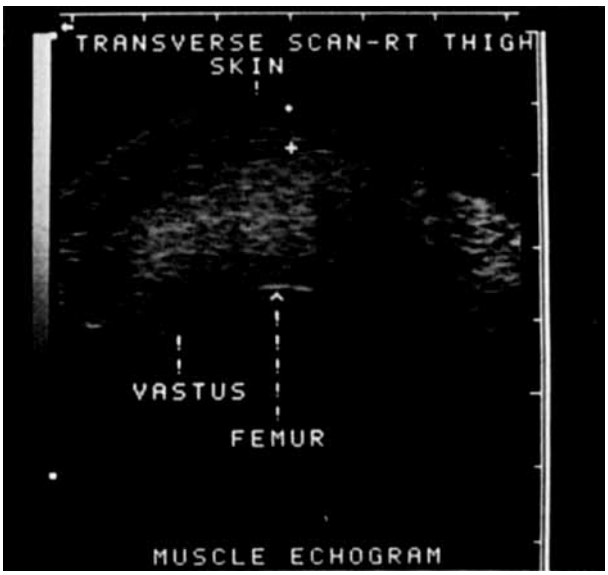

FIGURE 4. Transverse scan of thigh in an 8-year-old child with progressive muscular dystrophy showing markedly increased muscle echogenicity without delineation of muscle groups. Reduced bone echogenicity is also seen (grade III).

Infantile Spinal Muscular Atrophy

The 10 infants with a clinical diagnosis of infantile spinal muscular atrophy had marked hypotonia and areflexia. They had grade 1–3 muscle

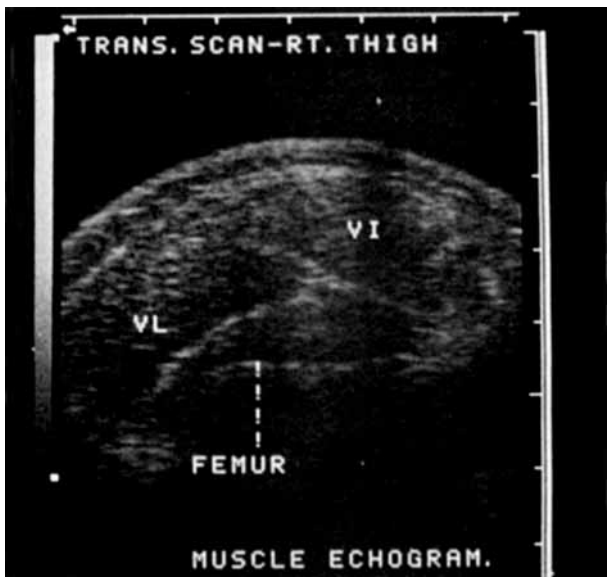

FIGURE 3. Transverse scan of thigh in a 5-year-old boy with progressive muscular dystrophy showing increased muscle echogenicity with delineation of muscle groups and visible bone echo. Subcutaneous space normal (grade II). VI, vastus intermedius; VL, vastus lateralis.

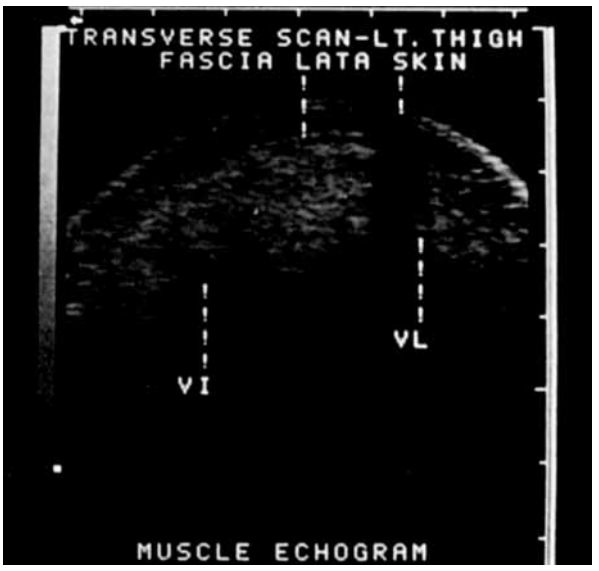

FIGURE 5. Transverse scan of thigh in a 7-year-old child with progressive muscular dystrophy showing marked muscle echogenicity with complete loss of bone echo (grade IV). VI, vastus intermedius; VL, vastus lateralis.

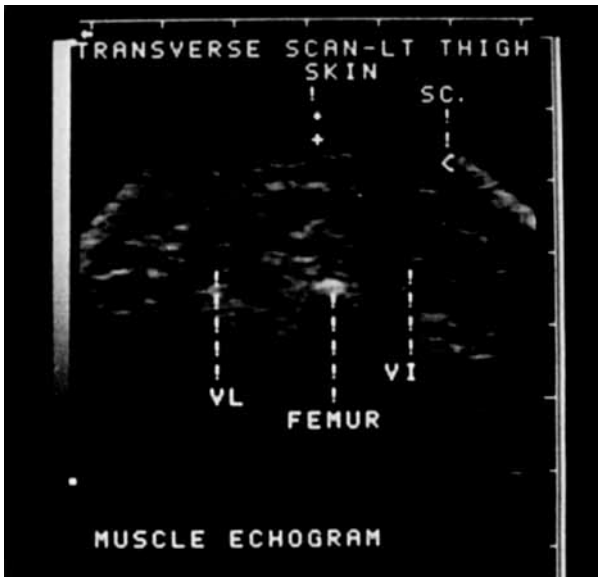

FIGURE 6. Transverse scan of thigh in a 6-month-old infant with suspected spinal muscular atrophy showing a marked increase in subcutaneous (SC) space, increased muscle echogenicity and well-preserved bone echogenicity. VI, vastus intermedius; VL, vastus lateralis; SC, subcutaneous space.

power. None had visceromegaly or cardiomegaly. One child was slow in sucking and cried feebly. Their serum creatine kinase values were normal. The electromyogram was normal in four cases and showed a neurogenic pattern in six cases. The ultrasonic scan of the thigh was abnormal in all cases. The bone echogenicity was normal, but the muscle echogenicity was increased. The depth of the subcutaneous space was also increased (Figure 6). The delineation of muscle groups was possible in most cases. In one case with well-defined bone echogenicity and increased muscle echogenicity, the depth of subcutaneous space was not increased. In another case, the bone echogenicity was decreased, with an increased subcutaneous depth and muscle echogenicity.

DISCUSSION

Our study has shown that ultrasonic scanning helps in the identification of cases of neuromuscular problems in children. This noninvasive procedure obviates the need for muscle biopsy in many children. An increased echogenicity of the muscle with varying changes in other parameters was consistently obtained in the two groups of children with muscle disease subjected to this test. There was a close correlation between ultrasonographic findings and muscle pathology in our cases of muscular dystrophy.

We found that delineation of the vastus intermedius and vastus lateralis is also an indicator of muscle involvement, as it was lost when the severity of the pathologic process increased. The muscle bulk was retained in the muscular dystrophy group despite degeneration, whereas there was marked atrophy in the spinal muscular atrophy group, as evidenced by an increase in the subcutaneous space in muscle sonograms. An occasional scan in a case of muscular dystrophy that shows increased depth of the subcutaneous space or an otherwise typical case of spinal muscular atrophy with no increase in subcutaneous space on the scan may be confusing. The ambiguous findings on scan may arise from different stages in the pathologic process or an overlap between the myopathic process and long-standing neurogenic pathology. Since it is a simple and noninvasive procedure, ultrasound may be repeated periodically for clarification in cases of doubt. Muscle biopsy may be resorted to if the ultrasonographic findings are equivocal. Ultrasonic scanning helps in detecting unequivocal muscle changes in clinically mild cases where there is apparently normal function. Although computerized tomography can be used to detect neuromuscular disorders,[6] ultrasonography is undoubtedly more convenient, less costly, noninvasive, and quicker in detecting neuromuscular problems in children.

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