THE SURGICAL MANAGEMENT OF SPINAL DEFORMITIES IN DUCHENNE MUSCULAR DYSTROHPY

S. Cervellati, P. Parisini, M. Palmisani, N. Bettini, R. Savini

The Authors evaluated the results of surgical management in 7 patients with spinal deformities due to Duchenne muscular distrophy. Or the basis of the results obtained and given the knowledge as to the natural history of the disease and correlated deformities, the Authors maintain that surgery still remains the only therapeutic means available capable of arresting the evolution of spinal deformity.

Key Words : Duchenne, spinal deformities, surgical management.

Spinal deformities in Duchenne muscular distrophy (DMD), favoured by the forced sitting position, usually appear between 13 and 15 years of age. As well as the sitting position, numerous other mechanisms have been cited as being responsible for this deformity. According to Gibson (4) and Wilkins (10), the main cause is the paralysis of the extensor muscles; according to Rideau (7), Siegel (8-9) and Dubousset (3), it is to be identified in the sloping of the pelvis, due to the asymmetry of the scar retractions in the region of the lower limbs. Ledoux (6) judges the cause of scoliosis to be the preferential position maintained by the wheelchair-bound patient in order to free and use to dominant hand, thus determining a permanent deviation of the spine.

For prognostic purposes, the evaluation of vital capacity is essential : Rideau (7) has divided Duchenne patients into three groups on the basis of V.C. values. According to this author, the vital capacity increases up to an age of 10-12 years, reaching a certain "plateau", and is then subject to gradual decline in subsequent years. A knowledge of the V.C. value in this "plateau" phase permits a prognosis of the restrictive pulmonary syndrome. The first type (V.C. < 1200 ml) is characterizied by a severe prognosis. The deformity is serious and advances rapidly with death occurring before 18 years af age. The second type (V.C. 1200-1700 ml.) is also characterizied by a serious prognosis with death occuring between 18 and 22 years af age. The third type (V.C. > 1700 ml.) has a better prognosis. The spinal deformity appears later and progresses more slowly. These patients have a longer life expectancy (20 - 25 years). In patients with DMD, spinal deformities hardly ever appear until the child is able to walk (8); thus, only in very rare cases do they appear before 11 years of age. The appearance of a spinal deformity is inevitable in all cases of DMD and, once it sets in, the deterioration is extremely rapid, seriously affecting the capacity of sitting without supports, with further damage to the respiratory function already precarious on account of the disease. There is no orthopedic corset able to impede the advancement of the deformity. It is significant that those authors-Gibson 1975 (4) and 1978 (5), Drennan 1979 (1) and 1984 (2) - who were once advocates of orthopedic treatment, eventually reached the conclusion that the prevention of deterioration of spinal deformity in DMD must be entrusted to surgical management.

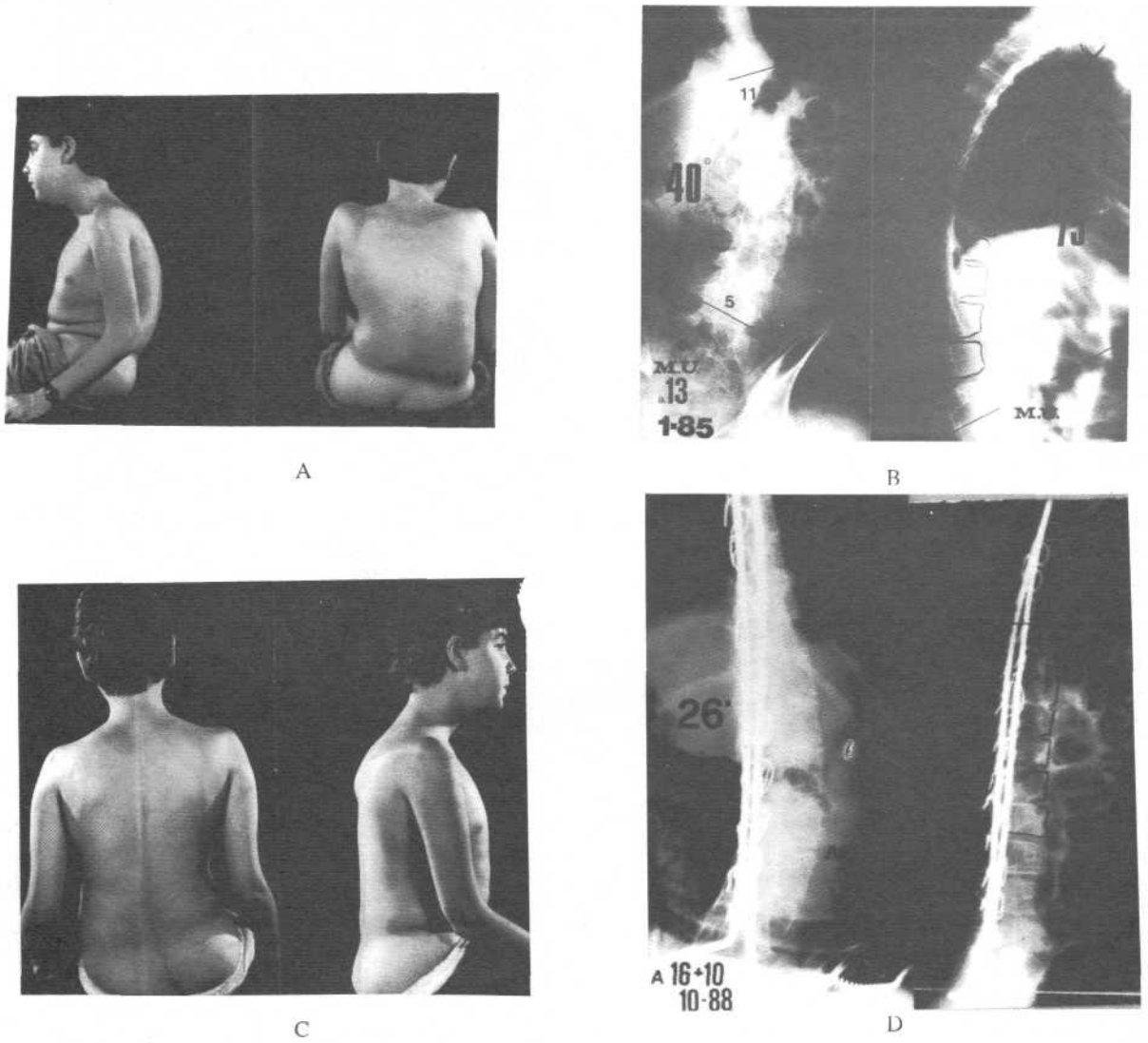
In recent years, thanks to a better knowledge of the natural history of the spinal deformity and the availability of safer surgical techniques, the indication to surgery has assumed the role of prevention in deformities of sure deterioration. Therefore, surgery is recommended today in cases where the patient is young, the vital capacity (> 50 %) permits surgery, the scoliosis is less than 30° and where serious cardial alterations are absent.

MATERIALS AND METHODS

Between 1985 and 1988, at the Scoliosis Centre of the Rizzoli orthopedic Institute in Bologna, we surgically treated 9 cases of spinal deformity in patients with Duchenne Muscular Distrophy. The diagnosis was made on the basis of family history and clinical, hematochemical, electromyographic and bioptic data.

S. Cervellati, P. Parisini, M. Palmisani,
N. Bettini, R. Savini : Scoliosis Centre Rizzoli Orthtopaedic Institute - BOLOGNA - ITALY

present in four patients.



A B C D

Fig.1.: 13 - year-old patient with kyphoscoliosis in DMD who has not walked for a year. This was the first patient we operated with spinal deformity in DMD (A-B). The control performed at nearly 4 years after surgery showed an unsatisfactory modelling of the bars along the sagittal plane and a partial relapse of the pelvic obliquity notwithstanding a good lumbarsacral arthordesis (C-D).

The average age was 13 years. All patients had lost the capacity to walk at a mean age of 3 years. All cases presented scoliosis with a mean angular value of 42° (10°-80°), & cases had associated kyphosis with a mean value of 42° (21°-75°); in 2 cases lordosis was present, with a mean angular value of 112° (90°-135°); 5 cases presented a mean pelvic obliquity of 14° (11°-18°). The preoperative vital capacity was equal to 1842 ml. (1250-2500). Echocardiographical alterations were

In all cases, a postcrovertebral arthrodesis was performed with Luque instrumentation. In 8 cases, the arthrodesis extented from the first three thoracic vertebra to the sacrum and in only one case from C7 to the sacrum. The operation had a mean duration of two and a half hours (1.40h-3.30h). Blood loss was 1200 ml. (700-1500 ml). In three cases, erythrocytes were recuperated during surgery, to be transfused during the post-operative phase. In one case only the post-

operative phase was passed in the intensive care unit for a period of two days. After a radiographic control at six/seven days subsequent to surgery, the patient was made to sit with no external supports.

RESULTS

The results refer to the first 7 patients treated with a mean F.U. of 2 years and 6 months (minimum 1 year, maximum 4 years and 3 months). The mean angular value of the scoliosis, 43° (10°-80°) before surgery, turned out to be 15° (0°-43°) in the postoperative period with a correction equal to 66 %. At follow-up controls, the angular value of the scoliosis had remained stable in 5 cases while in 2 cases a mean loss of correction of 12° (11°-13°) was recorded, although the instrumentation had not yielded.

A pathological thoracolumbar kyphosis with a mean angular value of 52° (41°-75°) was present in 5 cases, which was corrected to 8° (0°-26°) with a correction of 85 %, stable at the follow-up control.

The correction of the hyperlordosis, present in 2 cases, was equal to 54 % : from 112° (90°-135°) to 52° (45°-60°), unvaried at follow-up.

The pelvic obliquity present in 3 cases with a mean angular value of 15° (11°-18°) had been completely corrected in all cases. The correction of the pelvic obliquity remained stable in two cases, while in one case, a relapse occurred at a distance of 3 years and 9 months after surgery, without any radiographic evidence of pseudarthrosis. This was the only complication recorded (Fig.l).

The vital capacity which presented preoperative values of 1626 ml. (1250-2100), remained more or less unvaried in the postoperative period : 1548 ml. (1250-2100); after a year's interval the mean value was equal to 1402 (1080-1740), stabilizing after a period of 3 years at slightly lower levels; 1396 ml. (990-1740).

CONCLUSIONS

Our knowledge of the natural history of the disease and the correlated spinal deformities (i.e. a brief life-expectancy, deterioration of the spinal deformity impinging on the vital capacity regardless of the type of orthopaedic corset used), suggests that surgery should be performed before the deformity becomes serious and at a time when the child is in a better condition to withstand the operation. Today, the indication to surgery should be collocated at between 10 and 12 years of age in patients with V.C. not less than 40 % - 50 % of normal values and a scoliosis not exceeding 30° : In such conditions, the operation is easier, more rapid and therefore involves less risk. The Luque technique, with or without arthrodesis, is still the preferred method in that it allows a satisfactory correction of the deformity : rebalancing of the trunk and pelvis, comfortable sitting position, the use of the arms to perform normal activities, rather than for support, without requiring any form of postoperative constriction. The instrumentation must involve the entire column, from the first or second thoracic vertebra to the sacrum. The bars must always be modelled on the sagittal plane in order to permit a correct sitting position. We performed arthrodesis (7) in the correction of spinal deformities in DMD. However, we believe that if the decision is made to operate a very slight deformity at less than 11-12 years of age, this method represents a valid solution with the unquestionable advantage of shortening surgery time and not impeding growth of the spine. Finally, it is no longer admissible to maintain a renunciatory attitude when dealing with spinal deformities in DMD. The knowledge of the natural evolution of the disease and the safer surgical and anaesthesiological techniques should induce us to use prompt surgery, able to prevent the inevitable deterioration of the deformity and to render the lives of these patients less desperate.

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