walking function, development of evolutive scoliosis and progressive decline of the respiratory function leading patients to premature death.

In 1985 Y. Rideau in France carried out a new global therapeutic strategy for treatment of lower limb deformities, scoliosis deformity and progressive restrictive syndrome.

The indication for surgery at the lower limbs is made very early, at the onset of the first signs of disease. The procedures are carried out at the same time and always bilaterally; they include: (i) hip section of superficial flexors; (ii) iliotibial band resection; (iii) subcutaneous tenotomy of semitendineous and gracilis; (iv) subcutaneous lengthening of Achilles tendons.

In the post-operative period, the patient begins exercises of active and passive mobility in few days and after three weeks recovers his performances; ambulation will remain almost normal for several years. A comparison of two groups of patients, the first precociously operated on the lower limbs, the other one not operated, shows better performances in the operated group.

The indications for surgical treatment of Duchenne scoliosis must be made after the loss of ambulation and not too late, to avoid the concurrent respiratory restrictive syndrome makes the patient inoperable. Over ten years ago, in Poitiers, a specific instrumentation for Duchenne scoliosis was created, providing for cylindrical rods fixed by peduncular screws at the sacro-lumbar level. On the dorso-lumbar level, the rod becomes flat to allow more flexibility of the trunk. The complications observed in a group of 55 patients operated for scoliosis, consisted in 2 cases of breaking of rods and 1 superficial infection. The surgery approach in DMD has the double aim to prolong the time of the autonomous ambulation and to avoid the evolution of scoliosis, limiting the harmful effects of the scoliosis on the respiratory function.

However, the surgery alone is unable to prolong the life expectancy in these patients, without treating the restrictive respiratory syndrome, first by nasal ventilation and then by elective tracheotomy, essential for the survival of the patient.

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Early indicators of respiratory impairment in muscular dystrophies: from bioengineering to rehabilitation

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Respiratory rehabilitation in Duchenne Muscular Dystrophy

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Lung function in Duchenne Muscular Dystrophy (DMD) is complicated by a decline in vital capacity (VC). After an initial period of normal increase in VC, that typically reach a plateau between 12 and 16 years, there is a period of decline, estimated in 200ml/year. The deterioration of the respiratory function (restrictive syndrome) is one of the two major causes of death in these patients and a close correlation between the development of respiratory symptoms and VC has been observed. An indirect consequence of intercostals muscles (IM) weakness is a reduction in respiratory system compliance (RSC), the ratio of the change inspiratory system volume to a change in pressure (RSC = AV/AP). Mobilisation of secretions, by postural drainage, is an important step in the clinical pathway of airway clearance and of vital importance in DMD. Therapy should be provided for no longer than necessary to obtain the desired therapeutic results. The use of airway clearance techniques, including assisted coughing techniques, both manual and mechanical, is strongly recommended. These techniques should always be included in the treatment of chronic NMD patients. Cough can be assisted by manual and mechanical means. All methods require a combination of improved insufflation of the lungs to achieve sufficient lung volumes for an effective cough in conjunction with adequate forced expiratory techniques to increase the patient’s natural, but weakened, cough. Glossopharyngeal breathing (GPB) is the act of the glottis taking air and propelling it into the lungs. Some authors affirm that GPB can sustain normal ventilation throughout daytime hours without using a ventilator, and safely in the event of ventilator failure during sleep in patients with reduced inspiratory muscle function. In our opinion there is not enough evidence for generalised use of GPB alone to avoid invasive ventilatory assistance.

The cough assist machine (the mechanical insufflator-exsufflator) has proven to be useful to airway clearance in patients with neuromuscular weakness. It provides both an inspiratory phase (to inflate the lungs) and an expiratory phase (for the actual cough), all in one piece of equipment. Studies have shown that the Cough-Assist machine is well tolerated, without increased risk for complications such as pneumo-thorax, gastro-esophageal reflux, or pulmonary hemorrhage. It also can be used through various interfaces such as mouth, full face mask, or endotracheal or tracheostomy tube.

The indications for tracheostomy include: 1) NIPPV no longer effective at treating hypoventilation; 2) NIPPV not tolerated by the patient; 3) Excessive oral secretions; 4) Resources for outpatient management with NIPPV not available in the community; 5) Failure to extubate patient who has been intubated for whatever reason. In our experience tracheostomy is performed in election, when the vital capacity is about 700-1000 ml, so that the patient can long remain without constant ventilatory support.