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

In 1985 Y. Rideau in France carried out a new global thera-
peutic strategy for treatment of lower limb deformities, scoliosis
defority 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 in-
clude: (i) hip section of superficial flexors; (ii) iliobial 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 re-
covers 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 scolio-

sis 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 in-
strumentation 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 ap-
proach in DMD has the double aim to prolong the time of the
operative phase (to inflate the lungs) and an expiratory phase (for
sufflator) has proven to be useful to airway clearance in patients
with neuromuscular weakness. It provides both an inspira-
tory 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, gas-
tro-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 toler-
ated 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 intu-
bated 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.

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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 develop-
ment 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 pres-
sure (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 ther-
apeutic results. The use of airway clearance techniques, includ-
ing 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 re-
quire a combination of improved insufflation of the lungs to
achieve sufficient lung volumes for an effective cough in con-
junction 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 propel-
ing 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-ex-
sufflator) has proven to be useful to airway clearance in patients
with neuromuscular weakness. It provides both an inspira-
tory 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
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