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## ABSTRACT

This paper describes the design, bio-mechanics, and program being utilized during the past two years for improved seating and spine support for boys with Duchenne Muscular Dystrophy. It relates these factors to quality-of-life issues for the boy and his family.

Perhaps the most certain thing we can say about the development of spine deformity in boys with Duchenne Muscular Dystrophy (including the Becker variant) is that it is highly variable. The pattern may be hyperlordosis, hyperkyphosis, scoliosis, or a combination. While some children will experience an early onset of a spine deformity which progresses very rapidly to a very severe curve, others will acquire only mild, insignificant spine deformity, and still others will appear throughout the spectrum between those two extremes.

There is a lack of precise definitions and numbers regarding the incidence of significant spine deformity in Duchenne M.D., but we can say that a majority (perhaps higher than 85%) of those boys will develop a spine deformity having a large impact on the quality of life for child and parent. Of the deformity patterns we see in this group, those that include large scoliosis curves are the most debilitating. The scoliosis almost always includes a lateral obliquity (tilted laterally) of the pelvis.

The reasons a spine deformity has a negative impact on quality of life are:

1. The trunk progressively loses the stability and posture necessary for independent function of the hands.
2. The sitting position becomes progressively more painful for at least two reasons. Pelvic lateral obliquity causes most of the upper body weight to be borne by only one side of the pelvis. As the scoliotic collapse becomes severe, the lower ribs on the concave side are driven downward against the iliac crest.

3. Distortion of the thorax can eventually compromise heart/lung space and function.
4. As the youngster loses independent hand function and sitting comfort, more burden falls on the family to help with his function and to make constant positioning adjustments for comfort.

It is of fundamental importance, as we treat these kids, that we keep these quality of life issues rather than the orthopedic deformity, per se, uppermost in our minds. For instance, a semi-reclined sitting position will compromise function for these boys more quickly and surely than the spine deformity that position was meant to prevent. The seating and spine support program, whatever it is, must, in a comprehensive sense, help the boy and family to have a greater quality of life.

In the past, virtually all known efforts to orthotically control spine deformity in boys with Duchenne Muscular Dystrophy have failed in the long range. Either they failed outright to achieve the orthopedic goal, they failed to effectively address the quality of life issues, and/or they were excessively technically complicated in their application. At the present time, in some parts of North America, the pendulum is swinging toward early spine fusions.

At Gillette Children's Hospital, we have been seating children with Duchenne (and Becker variant) Muscular Dystrophy for about nine years. Although we were learning much about the nature of the deformity, about the function of these children and their families, and about how to work with them, progress seemed slow until two years ago. In early 1983, we combined the use of a simple soft abdomino-thoracic corset with the Gillette Sitting Support Orthosis (see figure 1). We eliminated all anterior restraints above the lap belt (Previously, we had used vests, panels, or straps to hold the boys back into the supportive plastic shell.). We continued to teach and stress the importance of "pelvic leveling" (elimination or minimizing of pelvic lateral obliquity) each time the child is put into the seat. We feel this improved seating program has been, thus far, extremely



Figure 1

successful. It appears to us that some key biomechanical factors have been addressed in a way that is consistent with improvement of the child's function and other quality-of-life issues.

The normal human torso receives its stability partly from the spinal column acting as a controlled stack of compression elements and partly from a multitude of muscles acting in several different ways. The paraspinal muscles have a direct action on the configuration of the spine; extending it, laterally flexing it, or rotating it. The abdominal (and to some extent the costal) muscles affect the stability and configuration indirectly, but very importantly, through their action on the abdomino-thoracic contents. Muscle action to constrict and control the circumference of the abdomen and thorax allow compressive body weight loads to be taken partly down through the fluid filled abdomino-thoracic cylinder rather than all acting down through the spinal column. This adds tremendously to the stability of the torso. Experiments on a cadaver spine have shown that when a spinal column is stripped of stabilizing musculature, it will buckle (like any flexible elastic column) under a compressive loading of only a few kilograms. As Duchenne Muscular Dystrophy progresses, destroying the effectiveness of the various muscle groups giving stability to the spine, it proceeds to collapse as would any elastic column being acted on by supercritical axial compression loads. The scoliosis pattern we see on x-ray is similar in configuration to the buckling pattern displayed by an elastic column pin joint (allowing

tilt) at its top. This biomechanical similarity would be of little practical value except for the fact that the stability of an elastic column under axial compression loads is strongly affected by the nature of the constraint at its end. Constraining the bottom of the column so that it cannot tilt, will roughly double the load carrying capability of the column (see figure 2).

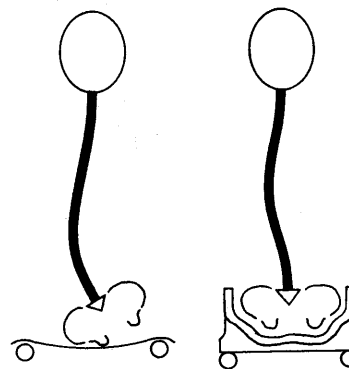
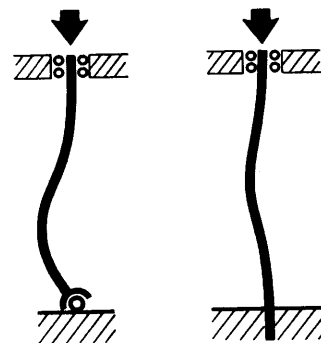


Figure 2

The Sitting Support Orthosis and soft corset combination increase spine/torso stability in three ways.

1. The soft corset substitutes for non-functional abdominal and costal muscles in a comfortable, unobtrusive way. It allows the compressive loads of body weight to bypass the spinal column by the biomechanism described earlier.
2. The SSO conforms to the pelvic portion of the anatomy and constrains it to remain in whatever orientation it is positioned in. Whoever puts the youngster into his SSO pulls up on the trousers on the low side of the pelvis and pushes down in the high side to slide the pelvis into a level (or at least minimally oblique) orientation. This is similar to increasing the

bottom end constraint on a flexible column to increase stability. Figures 3 and 4 show x-rays taken just before and after the pelvic leveling procedure was performed on one patient. The Cobb angle is reduced from  $36^{\circ}$  to  $20^{\circ}$  just by this quick procedure performed when positioning the child in his SSO. This patient was not wearing a corset for either x-ray. Please note that once a spine deformity has progressed to a rather severe degree, the pelvis can be leveled only to the degree that the deformity is flexible. We have come, over the years, to see frontal plane pelvic alignment as the most important variable in seating the neuromuscularly impaired youngster with scoliosis.

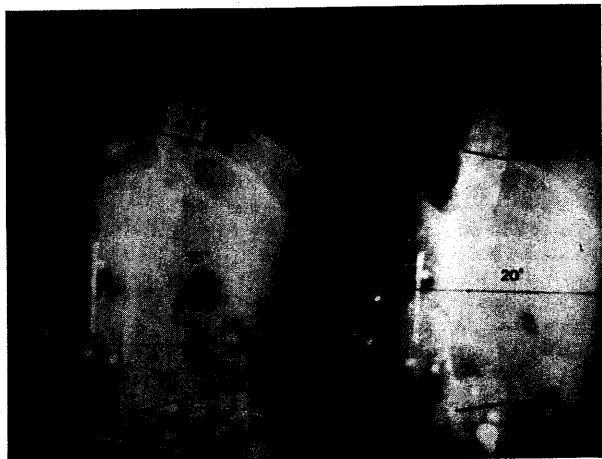


Figure 3

Figure 4

Pelvic control and orientation in the frontal plane relates not only to spine stability and all that derives from that; it relates also to the uniformity of pressures in weight bearing areas and alleviating the progressive deterioration of sitting comfort referred to earlier.

3. The upper portions of the SSO shell serves as props to prevent the torso, as a unit, from toppling backward, right or left. Forward position of the torso is controlled by the boy per his functional needs.

The corset, when properly adjusted, will also help the diaphragm to have a greater excursion making deeper inspirations possible. Some caution must be exercised. If the corset were adjusted extremely tight, it could compromise pulmonary function by preventing diaphragm descent. It is not difficult to avoid this unlikely complication.

It is the intent of the program here at Gillette to begin the scoliosis control program while the curve is still small

(between  $20^{\circ}$  and  $30^{\circ}$ ). However, some youths enter the program from other clinics with much larger curves. Figures 5 and 6 show same day x-rays with and without the SSO/corset combination (the SSO/corset x-ray includes pelvic leveling). In this case the pelvic lateral obliquity is reduced from  $30^{\circ}$  to  $14^{\circ}$  and a  $65^{\circ}$  scoliosis was reduced to  $35^{\circ}$ . Curve control of this magnitude is not unusual as long as the deformity is still very flexible.

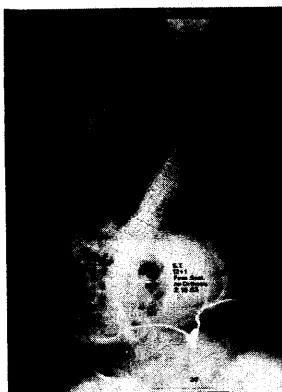


Figure 5



Figure 6

More gratifying than the x-ray evidence is the youngsters reaction. Their comfort, increased sitting height, better sitting stability, and deeper breathing combine to give them a feeling of increased well-being. Their function is maintained or increased, and they feel encouraged.

It must be emphasized that devices do not solve problems of this nature. Only a program including the following things will be successful:

1. Comprehensive education of the family as to the reasons for the prescription and the bio-mechanical principles.
2. Attention to finding and solving the functional and mobility problems related to seating before they go home with the new equipment.
3. Provide effective equipment and show the family evidence (i.e. x-rays) of its effectiveness.
4. Follow-up to solve growth, function and comfort problems as they occur and to monitor effectiveness.

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