Occlusal characteristics of children with spinal muscular atrophy

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Introduction

The occlusal status of children with neuromuscular disease remains poorly understood. Muscular dystrophy, especially Duchenne type muscular dystrophy, has been best described.[1-8] Children with myotonic and Duchenne dystrophies have greater risk of developing malocclusion due to weak or incompetent orofacial muscles.[1-3, 5, 8-10]

This study evaluates the occlusal characteristics of children with spinal muscular atrophy (SMA). SMA, with a prevalence of 1 in 25,000 live births, is the second most common neuromuscular disease of childhood following the muscular dystrophies.[11] It is usually inherited as an autosomal recessive trait, although sex linkage and dominant forms also have been described. The severity of the disease determines which muscles are affected. The proximal limb muscles usually are affected more than the distal muscles and the lower limbs more than the upper. Muscle weakness and atrophy have been attributed to anterior horn cell degeneration, while bulbar symptoms result from involvement of the brainstem motor nuclei in some instances. To date, the occlusal characteristics of children with SMA have not been described.

Methods and materials

A cross-sectional sample of 24 patients (10 males and 14 females) diagnosed with SMA was selected from active patients at the Texas Scottish Rite Hospital for Children, Dallas, Texas, and Children's Hospital, Cincinnati, Ohio. Data were collected over a 1.5-year period between 1990 and 1991. The mean age of the sample was 9.4 years; with seven subjects younger than 8 years old, nine subjects between 8 and 10 years old, and eight subjects older than 10 years old. None of the children had extractions without space maintenance, orthodontics of any kind, body braces, or mouth pieces for communication. Diagnostic criteria included the following:

1. Onset of weakness before age 18 years
2. Evidence of denervation by electromyogram or muscle biopsy
3. Normal nerve conduction velocity
4. No sensory deficiency on neurological examination.

The Treatment Priority Index (TPI) was chosen to assess occlusion because reliable reference data are available for comparison[12-13] and because the index is more replicable among different observers than clinical judgment based simply on experience.[14] All assessments were performed by one operator. The TPI summarized the following five components:

1. Buccal segment relations—describe the anterior-posterior relationship of the mandibular with the maxillary teeth (based most often on the first molars)
2. Overjet—measured with a Boley gauge as the distance from the most anterior labial surface of the permanent lower incisor to most anterior labial surface of the upper incisor
3. Overbite/openbite—measured in millimeters as the vertical overlap of the incisal edges or space between the incisal edges
4. Tooth displacement—based on a count of teeth rotated $\geq 45°$ and displaced ≥ 2 mm
5. Posterior crossbite—measure of buccal-lingual deviation in the canine and molar areas; it assumes the maxillary tooth has deviated.

Weights were assigned to each component and the sum of weights provides the individuals' TPI score, which represents the subject's overall measure of malocclusion. One-sample chi-square tests were used to compare the observed frequences of the SMA sample to the expected NCHS frequences. Only the 19 children younger than 12 years old were used for statistical comparisons with published reference data.

Results

Table 1 shows that the buccal segment relations of the SMA group have a significantly higher ($P = 0.02$) prevalence of distoclusion (64%) than the reference populations (39%). Since the occurrence of mesioclusion was slightly greater than previously reported for younger children, neutroclusion of SMA children is significantly less common than expected.

Overjet and overbite also showed significant differences between the SMA and reference groups. A greater percentage of SMA children had overjets of ≥ 7 mm. There was also an increased prevalence of anterior openbite (36%).

Children with SMA also have more rotated and displaced teeth than expected. Moderate to severe crowding—indicated by two or more teeth rotated $\geq 45°$ or displaced ≥ 2 mm—was evident for 64% of the SMA

Table 1. Relative frequency (%) of TPI components

	NCHS Age 6–11	NCHS Age 12–17	SMA All Ages (N = 24)
Buccal segment relationship*			
Neutroclusion	51.8	53.4	24.0
Distoclusion	38.8	33.6	64.0
Mesioclusion	9.4	13.0	12.0
Overjet (mm)†			
0–4	69.5	70.1	48.0
5–6	19.7	19.4	20.0
7–8	7.0	5.6	16.0
9+	2.7	2.8	16.0
Overbite/openbite (mm)†			
≤ 0.00	97.6	97.9	64.0
1–1.99	1.0	0.9	12.0
≥ 2.00	1.4	1.2	24.0
Tooth displacement score†			
0–2	82.9	29.2	36.0
3–4	10.3	18.8	20.0
5–6	4.0	18.4	20.0
7–8	1.9	13.3	12.0
> 8	0.9	20.2	12.0
Buccal crossbite (N of teeth)			
0	98.0	94.7	92.0
> 0	2.0	5.3	8.0
Lingual crossbite†			
0	91.9	88.3	56.0
1–2	5.1	8.9	20.0
> 2	3.0	2.8	24.0

Significant differences between observed and expected frequencies.
*$P < 0.05$. †$P < 0.01$.

sample. Almost 25% of the children had seven or more displaced teeth.

The prevalence of buccal crossbite among SMA children was expected, but the frequency of maxillary lingual crossbite of one or more teeth in the SMA group was significantly higher than expected. Approximately 44% of the SMA children had at least one posterior tooth in lingual crossbite; 24% had two or more teeth in lingual crossbite.

As expected, the TPI scores were significantly higher among SMA than control samples (Table 2). Differences are due to the much higher proportion of SMA children with severe (24%) and very severe (36%) malocclusions.

Discussion

This study, the first to evaluate the occlusal status of children with SMA, shows a much greater prevalence of malocclusion than for unaffected individuals. Approximately 60% of the children had malocclusion for which treatment is either highly desirable or mandatory. Their malocclusions are not simple; they are comprised of molar distoclusion, excessive overjet, openbite, crowding of the posterior segment, and maxillary collapse.

Two explanations for the observed differences are possible, depending on whether or not there is direct involvement of the orofacial muscles. Assuming that the orofacial muscles are not involved in children with SMA, as suggested by Merlini and coworkers,[15] the malocclusion might be attributed to mouth breathing. Respiratory difficulties associated with weak intercostal and diaphragmatic muscles have been implicated previously as important factors in determining fatalities of SMA children.[15] Moreover, some components of malocclusion described for the SMA sample are characteristic of habitual mouth breathers without muscle disease.[16]

Alternatively, the malocclusion might be related to weak masticatory and facial muscles. The incidence of malocclusion is associated with changes in dietary consistency,[17,18] and is also directly associated with masticatory stresses.[14] While no data for masticatory muscle function are available, subjects with SMA have been reported to have feeding difficulties[19] and to be unable to suck efficiently through a straw.[20]

Abnormal muscle function is a primary etiologic factor based on the similarity of malocclusion of this sample of SMA children and patients with muscular dystrophy. Reduced muscle function associated with myotonic dystrophy has been shown previously to have a profound effect on occlusion. Kiliaridis and coworkers[8] reported that 46% of patients with myotonic dystrophy had distoclusion, 38% had overjet, 33% had openbite, and 50% had unilateral or bilateral crossbite. Openbite, crossbite, and mouth breathing were also reported by Gazit and coworkers.[7] Subjects with

Table 2. Relative frequency (%) of treatment priority index*

TPI Score	NCHS Age 6–11	NCHS Age 12–17	SMA All Ages (N=24)
< 1	22.9	10.5	4.0
1–3	39.7	34.5	20.0
4–6	23.7	25.8	16.0
7–9	8.7	13.0	24.0
10+	5.0	16.2	36.0

* Significant differences between observed and expected frequencies ($P < 0.001$).

Duchenne dystrophy, which also involves the orofacial musculature, have been shown to have higher than expected prevalence of posterior crossbites and openbite.[1-3, 5, 6] Of 43 subjects with Duchenne dystrophy studied by Ghafari and coworkers,[10] 58% had posterior crossbite and 21% had anterior openbite.

The prevalence and degree of malocclusion of the SMA sample is significantly greater than for the U.S. reference population. We suggest that reduced masticatory and facial muscle forces are at least partially, if not primarily, responsible for the malocclusions observed. The observed arch constriction, with the associated rotated and crowded teeth, might be expected to increase the children's potential for dental disease and could compromise their nutrition. The relevance of orthodontic treatment of the children must be considered relative to their life span, potential for relapse, functional improvement, and resources required. Further studies are required prior to prolonged treatment of SMA children. A compromised treatment plan may include alleviating the crowding through extractions rather than complete orthodontic alignment of the dentition.

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