WHETHER OR NOT TO TREAT CHILDREN WITH DOWN SYNDROME

I discuss here what well-meaning orofacial myologists should know before considering whether to treat the OMDs seen in children with Down Syndrome. (You probably already know that there is no "s" after Down; that is, it is not Downs syndrome, but instead, Down Syndrome).

PRIMARY MORPHOLOGIC CHARACTERISTICS

This genetic syndrome (Trisomy 21 Syndrome) has many facial and oral morphological characteristics which are important to evaluate when considering treatment of the OMDs usually seen in individuals with this syndrome. The primary features of interest to orofacial myofunctional clinicians are: a small maxilla with maxillary retrusion, a small nasopharynx, an acute cranial base angle, an adenoid mass that often occludes the posterior entrance into the nose, a normal size tongue in a small oral cavity, varying levels of cortical functioning, and difficulty maintaining a nasal breathing pattern.

Let's go through these characterizing features individually: The **maxillary retrusion** seen in children with Down Syndrome is related to the maxilla being small in overall size. A small maxilla results in a reduction of the area of the nasal cavity that delimits establishing and maintaining a nasal method of breathing.

The **nasopharynx** is also small due to the retruded maxilla, but as well, the angle of the cranial base is usually somewhat acute in children with Down Syndrome, as well as being a characteristic of other midfacial retrusion syndromes. You can gain a perspective about the impact of an acute cranial base angle on the depth of the nasopharynx by observing the drawing below of a lateral view of the angulation changes seen in the cranial base. With an acute cranial base angle, the distance between the posterior wall of the pharynx and the posterior entrance into the nose (the posterior choanae) is diminished, thus narrowing the horizontal diameter of the nasopharynx and further hampers the ability of children with Down Syndrome to habituate a nasal mode of breathing.

As you know, the **adenoid mass**, when present, forms an attachment on the posterior wall of the pharynx, extending vertically to the base of the skull and laterally, may occasionally circle and close over the opening of the Eustachian tubes that are located on the lateral walls of the pharynx, resulting in many bouts of otitis media. The adenoid mass may impinge on the posterior opening into the nose (posterior choanae) and in doing so, can interfere with the flow of air in or out of the nose. The consequence of these upper airway interferences is that children with Down Syndrome will adapt by reposturing the tongue forward as a means of maintaining the airway, to make up for the reduction in size of the nasopharynx and nasal cavity. Mouth breathing is obligatory with most children with Down Syndrome.

Dr. Dan Subtelny, an orthodontist, described the **tongue** in individuals with Down Syndrome as a "relative macroglossia"; that is, children with Down Syndrome have a normal size tongue in a small oral cavity; accordingly, there is no true macroglossia present. The tongue appears macroglossic because it protrudes, but the mandible to which it is attached is normal in size and yet, the maxilla to which it also relates is small. The oral cavity is small because of the retruded and small maxilla, and the tongue naturally adapts to the small areas above by posturing forward.

With Down Syndrome, the normal size tongue and mandible, coupled with a small nasopharynx and maxilla, result in the tongue resting and functioning forward. As mentioned above, this protruded rest position helps to maintain the airway, and oral breathing is the result.

Individuals with Down Syndrome show a wide variation in cortical functioning ability. Those with reduced cortical functioning will often lack the ability to control the vertical movements of the tongue in therapy, replying instead on horizontally-directed tongue patterning. For such patients, substituting a tongue tip rest position at the lower incisors helps to provide a good starting position for the tongue for speech and swallowing therapy. Achieving this rest position depends on an ability to breathe normally with the tongue repositioned to a rest position at lower incisors. FYI - it is a myth that a tongue at lower incisors rest position leads to negative dental changes. (This myth needs to be eliminated among orofacial myofunctional clinicians, as it is unfounded and untrue).

Children with Down Syndrome often exhibit flaccid tongues that lead to a reduction in ability to perform well on oral diadochokinetic testing. When the starting position for speech is with the tongue protruded, producing vertical tongue movements is further compromised, as is the starting position for speech sound productions.

THE IMPORTANCE OF OBTAINING A LATERAL CEPH ON INDIVIDUALS WITH DOWN SYNDROME

Having said the above, if you contend that orofacial myofunctional therapy can be successful with children with Down Syndrome to: 1) retract a protruded tongue, 2) achieve a lips-together rest posture and 3) maintain a nasal pattern of breathing, within an environment of the delimiting morphologic factors identified and discussed above - *would it not be indicated to obtain (as a **minimum** diagnostic assessment) **a lateral cephalometric x-ray film** to evaluate the posterior airway, the size of the adenoids, (and the faucial tonsils that compete for the same space as the posterior tongue), and to also assess the nasal cavity and the ability of a child to breathe nasally?* I have yet to see any report of lateral cephalometric findings in children with Down Syndrome that have been recommended to undergo therapy.

POOR NASAL HYGIENE

It should also be remembered that since children are notorious for having poor oral hygiene, they also have poor *nasal hygiene*; that is, they do not do well in keeping their noses clean and free of debris which interferes with nasal breathing. It would be no surprise to find that children with Down syndrome would exhibit poor nasal hygiene that would further exacerbate the other upper airway interferences characteristic of individuals with Down Syndrome. It is no secret that most children have to be reminded to blow their noses, and many have to be taught how to perform this task.

INTRAORAL EXAMINATION

Among the considerations in performing therapy with children with Down Syndrome, an intraoral exam of the posterior airway is also recommended. Can the posterior pharyngeal wall be easily observed? How does the soft palate elevate, and does it do so normally, elevating up to the plane of the hard palate?

Is a child under consideration for therapy able to retract the tongue and sit with lips together? This task will not be possible with most children with Down Syndrome, however, with continued facial and pharyngeal growth extending into the teenage years, therapy might then be possible following vertical and horizontal growth expansion of the pharynx, maxilla, and the normal reduction in size (involution) of tonsils and adenoids that usually begins around ages 9-12 years.

OMD THERAPY FOR DOWN SYNDROME CHILDREN

For any advocates of early myofunctional therapy for children with Down Syndrome, I wonder on what basis that therapy is recommended? I also wonder what diagnostic information has been collected on such children that would encourage a clinician to attempt to reposition the tongue, achieving a nasal pattern of breathing with a lips-together rest posture? For most children with Down Syndrome, this would be impossible as per the delimiting morphological factors detailed above. The very minimum of diagnostic evidence to evaluate a child's candidacy for therapy with Down Syndrome would include a lateral ceph. **Is this done?** Most likely, no.

In the early 1980s, some well-meaning Japanese and Canadian clinicians decided separately that they would undertake a surgical tongue reduction in children with Down syndrome - to improve their appearance. The real motivation for surgery was from parents who were concerned about the appearance of their children. The parents also thought that tongue reduction surgery would improve speech. Such clinical experiments, done in good faith, failed for a variety of reasons including the finding that the tongue resumed its previous protruded position even after a surgical reduction in size.

Altogether, and according to the characteristics discussed here, the role of OMT with children with Down Syndrome is questionable and those who are providing and advocating for such therapy have not reported any long-term successes from any short-term successes claimed. In the teenage years, however, some individuals with Down Syndrome may become appropriate candidates for therapy following the additional pharyngeal and facial growth that has occurred. Some individuals may then be able to be taught a nasal pattern of breathing.

A few added thoughts: One of the worst things a clinician can do is under-estimate a child's ability. A rule to live by, that you hopefully already practice is that it is better to over-estimate a client's abilities and mental capability than to under-estimate and find out later that you were wrong. Such is the case with many teenagers and adults with Down Syndrome.

THERAPY PERSPECTIVES FOR INDIVIDUALS WITH DOWN SYNDROME

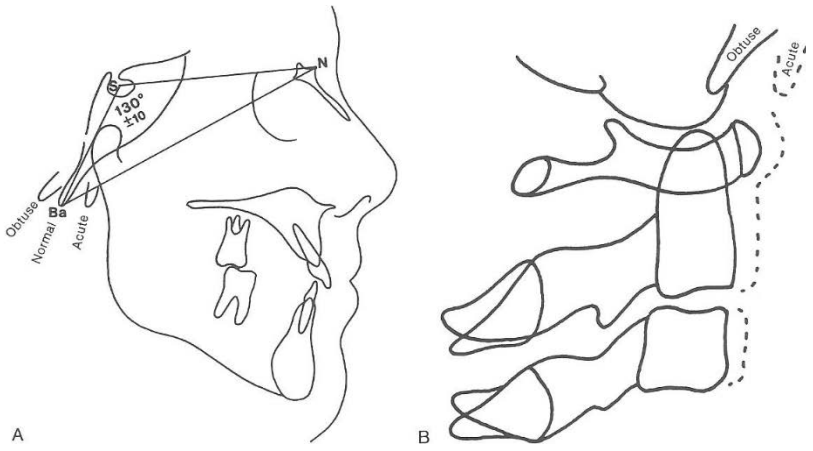
The development of a treatment strategy in treating OMDs in patients with Down Syndrome should involve a decision as to whether to work on the *deficits* that a client possesses, or work on *strengths* identified. Individuals with Down syndrome function well in the horizontal plane of space; they can easily protrude the tongue but have difficulties with controlled vertical movements of the tongue. With regard to the ability to protrude, a reasonable target for linguoalveolar sounds is the back of upper teeth rather than the alveolus. In some cases, using the lingual surfaces of lower incisors is an appropriate target, since /t/, /d/, /n/, /l/ can be made with acoustic acceptability with the tongue tip being released from a contact at the lower incisors. Sibilants are easily produced with the tongue tip at the lower incisors, with airflow passing over the blade of the tongue and then released between the upper and lower incisors that are slightly apart, thus creating the sibilance that defines these sounds. Also, all /r/ sounds can be produced acoustically normal with the tongue tip at or released from a lower incisors contact.

For older clients and teenagers, working to reduce the flaccidity of the lateral surfaces of the tongue may aid in repositioning the tongue at rest. As all children grow, and as the mandible also grows, teenage children with Down Syndrome may have the ability to retract the tongue at rest by adapting to growth changes that have occurred in the vertical plane. Those that are successful are seen on lateral cephalometric x-ray films to have adapted to a lower position for the hyoid bone. The hyoid is a good marker to assess tongue adaptation. Accordingly, some teenagers and adults with Down Syndrome are able to respond to exercises to achieve lip competence and assume a more appropriate, less protrusive tongue rest posture as growth occurs. Growth into the teenage years involves vertical, transverse, and horizontal growth of the jaws and vertical growth of the pharynx. This results in increased oropharyngeal room in the teen years for patients with Down syndrome to be taught more adaptive skills and oral postures that can result in improved facial appearance.

The key to choosing appropriate individuals with Down Syndrome for your therapy is *thorough evaluation* to identify those individuals who may benefit from your skills. I would insist on obtaining a lateral ceph in an orthodontic office and some help from a friendly orthodontist to evaluate the ceph regarding a child's candidacy for therapy, based partially on ceph findings related to the airway.

DIAGNOSTIC THERAPY

I also endorse *diagnostic therapy.* Whenever you are unsure of the value of therapy and want to try in spite of whatever reservations may be involved, it is recommended that if you proceed, you do so within the perspective of *diagnostic therapy*. In that way, you clearly identify the purpose of your work and that there may be delimiting factors that will cause you to cease and desist therapy at some point. In every case, you will certainly learn something.



CRANIAL BASE ANGULATION (THE PHARYNGEAL WALL IS LOCATED RELATIVE TO THE CRANIAL BASE ANGULATION)