In humans, abnormal masseteric contractions have also be demonstrated in the presence of mouth breathing [38], suggesting that abnormal orofacial muscle activity links nasal obstruction to deficits in structural airway growth. Secondary posture changes associated with chronic mouth breathing have also been identified [30,39,40]. Interestingly, in the Rhesus monkey model, removal of nasal obstruction at 6 months led to return of normal nasal breathing and yielded improved morphometric development, whereas continued impairment of normal nasal breathing led to continued mouth breathing and abnormal oral-facial growth and development.

Interactions Between Orofacial Function and Growth: Observations in Disorders Involving Upper Airway Muscle Dysfunction

Increased nasal resistance is unlikely to be the sole avenue to chronic oral breathing and subsequent craniofacial growth alterations. In humans, neuromuscular disorders provide further insight about the relationship between altered muscle tone and changes in craniofacial development. [41-43] for example, in the myotonic dystrophies and some congenital myopathies, abnormal orofacial muscle tone leads to impaired development of craniofacial structures. Presentation includes increased vertical facial growth, a narrower maxillary arch, and deeper palatal depths. In these disorders, abnormal orofacial muscle tone has consequences for the growth of upper airway structures, in association with early and chronic mouth breathing and frequent development of obstructive SDB, with rates reported to be 43-69%.

Ehlers-Danlos Syndrome (EDS), on the other hand, is an inherited connective tissue disorder involving abnormal collagen. The collagen-vascular mutations seen in Ehlers-Danlos syndrome lead to abnormal facial growth. These changes lead to narrow nasal passages, forcing mouth breathing, particularly during sleep [44]. Clinical evaluation demonstrates abnormally long facial shape, narrow and/or high maxillary hard palate, often with crossbite. While initially only abnormalities of the naso-maxillary complex maybe seen, as patients get older, defects of the mandibular condyle may become evident, which we hypothesizeis promoted by the presence of chronic oral breathing. A similar pattern of facial growth abnormality is noted with dental agenesis: Mutations in homeobox genes including those involved in normal tooth development (including those with ectodysplasin A -EDA- and WNT 10A genes as noted in our patients) lead to narrow facial skeleton, mouth breathing and, in our study, to SDB [45-48].

History of prematurity is another circumstance associated with higher likelihood of sleep disordered breathing in childhood, and is therefore another interesting example of the interplay between muscle tone, craniofacial growth, and nasal versus oral breathing route. Recently a large convenience cohort of 300 premature infants (36 to 27 weeks gestational age) was followed for 3 years after birth with clinical evaluation, psychometric testing, facial and oral dimension assessment, and PSGs at birth, 12, 24 and 36 months of age. [49-50] as expected, the infants had a variable degree of hypotonia, with severity generally related to degree of prematurity. High and narrow hard palate (HNP) was noted at birth in many premature infants and was more common with younger gestational age; HNP infants were more likely to exhibit mouth breathing; and their mean apnea-hypopnea index (AHI) was significantly higher compared to the non-high/narrow palate group; and the HNP infants were also found to have significantly more feeding difficulties. While many infants with feeding difficulties did not receive early feeding/orofacial education services, including sensory stimulation training and oral-facial exercises, 42infants did receive these services and rather remarkably, demonstrated improvements in palatal dimensions at 36 months relative to those without orofacial training. We hypothesizethat orofacial muscle development played a role in normalization of palatal structures at 36 months.

There were also 23 infants who had a normal palate at birth, but evolved toward HNP, mouth breathing and SDB, suggesting that postnatal developmental factors also alter palatal growth [49].

In summary, whether experimentally induced or developmentally provoked, science and nature have provided with examples of the interplay between increased nasal resistance and/or poor muscle tone leading to chronic oral breathing, and subsequent altered craniofacial dimensions. We believe that the presence of chronic oral breathing is both a marker of an inadequate or obstructed nasal-pharyngeal airway, and a marker of persisting abnormalities in the developmental interplay between muscular control, breathing route, and structural growth of the upper airway.

Applications in the Treatment of Pediatric Sleep-Disordered Breathing

While the above considerations are suggestive, much more work is needed to understand chronic mouth breathing as a marker of, and possible precipitator of, SDB in pediatrics. To further understand the proposed detrimental role of abnormal orofacial tone and mouth breathing during sleep, PSGs of 64 non-obese children aged 3 to 9 years (with mean AHI=8.5 events/hour and mean flow limitation= 76%), and who had PSGs pre- and post- treatment for SDB, were assessed [51]. In our lab, an oral-only sensor (utilizing an oral scoop) is used toaccurately and simply monitors mouth breathing [52]. In all of the baseline PSGs of the 64 children with SDB, there was evidence of excessive mouth breathing (defined as at least one third of total sleep time) on baseline diagnostic PSG. After adenotonsillectomy, 26 children had an AHI equal or higher than 1.5 events/hour. These children continued to have evidence of significant oral breathing. An additional 9 children whose AHI was under 1.5 events per hour also continued to have oral breathing - this is a very interesting group deserving further study. Clinically, children with SDB and persistent chronic mouth breathing after T&A may be referred for myofunctional therapy [53] in addition to usual therapies (e.g., consideration of anti-inflammatory medications, rapid maxillary expansion, CPAP). Eighteen children returned for 12 month follow-up, with only 9 having completed 6 months of myofunctional therapy. Though the numbers are very small, those who completed myofunctional therapy in addition to usual therapies were observed to have had improvements in nasal breathing as well as sleep, as measured by AHI and nasal flow limitation, beyond improvements seen in children without myofunctional therapy [51]. This suggests that even after nasal obstruction has been alleviated, improving muscle function of certain airway muscles, including the tongue, may improve function and/or growth of the upper airway, with resultant consequences for nasal breathing during sleep [51-55].

Observations and Conclusions: The Interplay Between Muscle Activity, Structural Growth, and Breathing During Wake and Sleep

The interaction between orofacial structural growth and muscle activity starts early in development, and the physiologic functions of suction, mastication, swallowing and nasal breathing in infancy play an important role in stimulating subsequent growth [55-58]. In the service of these functions, orofacial muscle use serves to help stimulate the direction and degree of growth. Mouth breathing is associated with altered oral-facial muscle activity and oral-facial growth. As such, its persistence is never normal. In fact, oral breathing has been termed "the most obvious manifestation of a syndromic pattern" involving a circuit of frequent infections, development of malocclusion, incorrect phonation, abnormalities of body posture, and changes in sleep. [30] Fortunately, oral breathing as a clinical sign has the advantage that its presence can be detected by simple direct observation, and its severity during sleep can be quantified with PSG.