MRI Comparison of Children's Vocal Tract Anatomy:

Poster 89

Houri K. Vorperian[1], Ray D. Kent [1, 2],
Waisman Center [1], Department of Communicative Disorders [2],
University of Wisconsin -

ABSTRACT: Comparison of preliminary data from magnetic resonance images (MRI) of 4 children with Down syndrome (DS) ages (mos;year) 0;10, 1;5, 3;2 and 8;10 and their age and percentile growth matched controls indicate that both groups have similar tongue length and area. However, children with DS have a smaller anterior oral cavity that does not accommodate a normal-sized tongue.

PURPOSE: Given the prevalence of speech sound errors (impaired or deviant articulation) in children with Down Syndrome (DS) [3, 4, 7, 8, 14, 15, 16, 28] the purpose of this study is to begin exploring the anatomic similarities and differences of the supralaryngeal speech apparatus in typically developing children and children with DS using Magnetic Resonance Imaging (MRI).

BACKGROUND: Some of the major physical features/characteristics of the head and neck for individuals with DS -- based mostly on observations -- that are considered to be of diagnostic significance include [1,17, 21, 22]:

- Head: Flattening the back of the head
- Face: Flat facial profile, slanting eyelids, depressed nasal bridge, smaller mouth and ears
- Tongue: Macroglossia (60% relative macroglossia)
- Palate: High and narrow palatal arc
- Dentition: Malocclusion (mandibular overjet, poterior cross bite)
- Feeding: DS infants are prone to have feeding problems [18]; and individuals with DS typically experience oral and pharyngeal dysphagia [26]
- Airway: Obstruction (acute and chronic); small airway, large tonsils/adenoid causing sleep apnea.
- Muscle tone: Decreased muscle tone, hypotonia and ligamentous laxity.

METHODS & PROCEDURES:

Subjects: Four children with Down Syndrome ages (mos;year) 0;10, 1;5, 3;2 and 8;10, and four age and percentile growth matched typically developing controls. Percentile growth was determined from growth charts specific for Trisomy 21[19] using weight rather than height (See figures 1 and 2). All subjects received magnetic resonance imaging (MRI) at the University Hospital for medical reasons that were judged not affect growth and development.

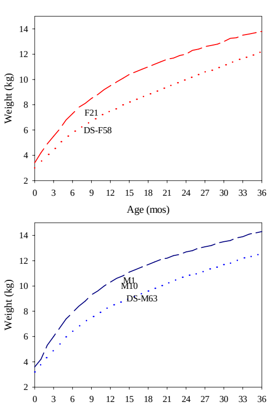


Figure 1. Growth curves for ages 0-36 months; dotted line represents DS growth curves, and dashed line typical growth curves for females (top) and males (bottom).

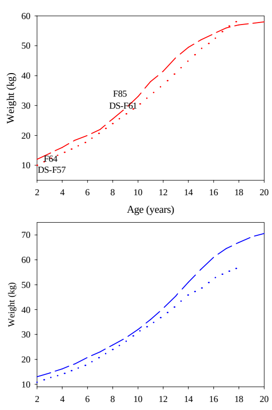


Figure 2. Growth curves for ages 2-20 years; dotted line represent DS growth curves, and dashed line typical growth curves for females (top) and males (bottom).

Procedures: Magnetic resonance images from children with Down Syndrome and their controls were measured using procedures we established in 1999 [30]. Using the specified anatomic landmarks (Figure 3), the following set of measurements were made in the head and neck region from midsagittal, parasagittal and axial slices.

Head length: (MSP-cm; g-op): The maximum distance from the glabella (g) to the opisthocranion (op).
Head height: (MSP-cm; n-v): The distance from the nasion (n) to the vertex (v).
Upper Facial Height (MSP-cm; n-sto): The distance from the nasion (n) to the stomion (sto).
Lower Facial Height (MSP-cm; sto-gn): The distance from the stomion (sto) to the gnathion (gn).
Facial Convexity (MSP-degrees; g-prn-pg or g-sn-pg): Theangular measure at the nose tip when the glabella (g) - pronasale (prn) - pogonion (pg) connect.
ANB Angle (MSP-degrees; sn-n-pg angle): Assesses maxillary growth or the maxillary to mandibular anteroposterior facial parameters.
Tongue Length (MSP-cm): The curvilinear distance along the dorsal superior contour of the tongue from tongue tip to valleculae.
Hard Palate Length (MSP-cm): The curvilinear distance along the hard palate contour from the anterior point of the incisor or tooth bud to the beginning of the soft palate.
Soft Palate Length (MSP-cm; soft palate + uvula): The curvilinear distance from the posterior edge of the hard palate to the inferior edge of the uvula.
Maxillary Arch Width (Axial-cm): The intercanine arch width at the buccal cusp surfaces (or dental buds in infants).
Maxillary Arch Length (Axial-cm): The distance of the median palatine suture of the hard palate measured from the anterior edge of the incisors' tooth bud or cusp, to the imaginary line drawn between the maxillary tuber.
Vocal Tract Length (MSP-cm): The curvilinear distance along the midline of the starting at the superior edge of the thyroid cartilage to the intersection with a line drawn tangentially to the lips.
Pharyngeal length: The curvilinear distance along the posterior pharyngeal wall above the soft palate extending from the nasal choana -- or posterior nares -- to the level of the thyroid cartilage or the end of the upper airway.
Mandibular Length and Depth (MSP & parasagittal plane-cm): The horizontal and vertical distances in the MSP from the mental protuberance to the orthogonal projection of the condylar process on the MSP.
Mandibular Width (Axial-cm): The distance between the temporal edges of the two condylar processes.

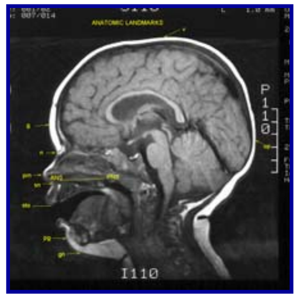


Figure 3. Anatomic landmarks. Defined in text. Midsagittal MRI slice of a male S (2;0)

Anatomic landmarks and reference lines used for measurements from the midsagittal plane were as follows:
ANS-PNS: The line drawn between the anterior nasal spine and the posterior nasal spine.
g : glabella - the most forward point on the forehead.
gn : gnathion - the lowest median landmark on the lower border of the mandible.
n : nasion - the point in the midline of the nasal root.
op : opisthocranion - the point situated in the occipital region of the head and most distant from the glabella.
pg : pogonion - the most anterior midpoint of the chin.
prn: pronasale - the most protruded point of the apex nasi.
sp : spinal point - the apex of the anterior nasal spine.
sto: stomion - the midpoint in the oral fissure when the lips are gently closed, with teeth shut in the natural position.
sn : subnasale - the point where the nasal septum and the surface of the upper lip meet.
Si : sulcus inferior -- or point B -- supramentale-- the most posterior midline point in the concavity between the infradentale –the highest anterior point on the gingiva between the mandibular central incisors-- and the pogonion.
v : vertex - the highest point of the crown of the head.

RESULTS: Preliminary data/measurements are presented on grouped bar graphs. The standard deviation of measurement error for soft tissue structures was smaller (σ= .12 ; n=74) than for hard tissue structures (σ= .20 ; n=79). Each figure -- bar graph -- is followed by a statement highlighting findings and where applicable followed by a statement comparing findings to reports by other researchers.

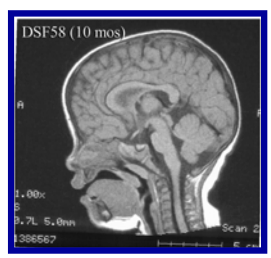


Figure 4. MRI of S DSF58 at age 10 mos

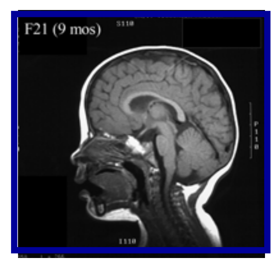


Figure 5. MRI of S F21 at age 9 mos

HEAD:

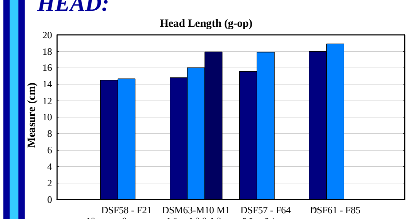


Figure 6 shows that head length (g-op) of the youngest DS subject DSF58 is very similar to her control F21, however, head length differences are present at all other ages.

Farkas et al. [9,10,11] report DS head circumference (a measure that correlates highly with head length) to be only 28.6% within normal measurement (within 1 to 2 SD from normally developing subjects). This percentage, however, is based on group data from 127 DS patients ages 7 months to 36 years.

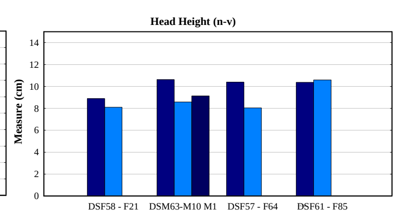


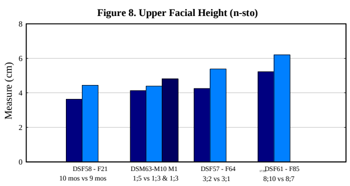
Figure 7 shows that head height (n-v) is typically greater for Ss with DS than their controls.

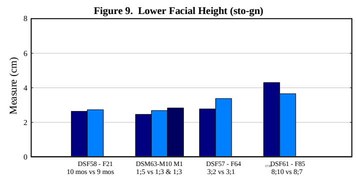
Farkas et al. [9,10, 11] report DS head height to be a measure that is 85% within normal measurements (within 1 SD of typically developing Ss). This is based on group data from 127 DS patients ages 7 months to 36 years.

Normal vs Down Syndrome -- Preliminary Data

and Lindell R. Gentry [3]
and Department of Radiology [3]
Madison

ASHA - Philadelphia, PA
November 20, 2004

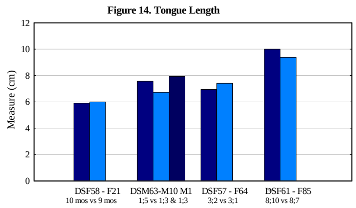




Figures 8 and 9 indicate that upper facial height is somewhat smaller in individuals with DS than their controls. Lower facial height, however, appears to be the same for both groups.

These results agree with Allanson et al.[1] who report DS subjects to have brachycephaly i.e. short-headed.

ORAL AND PHARYNGEAL CAVITY:



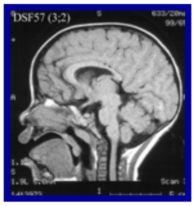


Figure 10. DSF57 at age 3;2

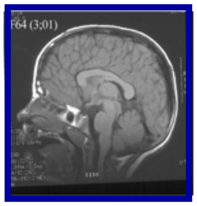
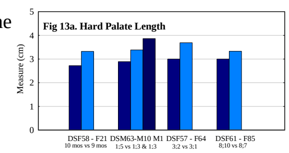


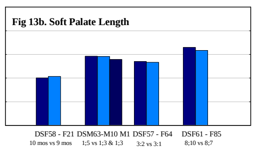
Figure 11. F64 at age 3;1

Figure 12. Tongue length in subjects with DS is similar to tongue length of their controls.

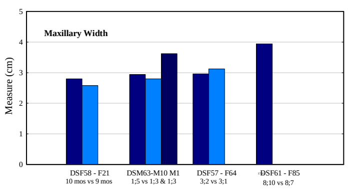
Krmpotic-nemanic & Schulz [13], Ardran, Harker & Kemp[2] and Uong et al.[29] report similar findings. Krmpotic-nemanic & Schulz use the term ***relative macroglossia*** which captures the essence of a smaller anterior oral cavity in DS subjects that does not accommodate a normal-sized tongue.

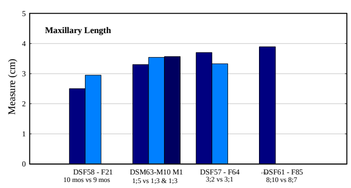
Figure 13 (ab) show that the hard palate is shorter in individuals with DS than typically developing subjects, but soft palate length is comparable.





These observations support findings by Uong et al [29] that hard palate length (nasal spine to end of palatine bone) is shorter in DS subjects, but soft palate length is similar to controls.





Figures 14 and 15 indicate DS subjects' maxillary width and length to be similar to their controls.

These observations, though contrary to descriptive physical characteristics of DS subjects, are supported by some researchers but not others. Redman et al. [23] and Shapiro et al. [26, 27] state that this nonmetrical impression of "narrow" and "high" palate is due to shelf-like palatal alveolar processes. Panchon-Ruiz et al.[20], however, report that the DS palate is not a reduced scale model of the typically developing palate. They report palatal morphology in DS to fit an elliptic paraboloid.

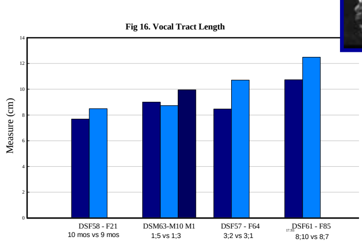


Figure 16. Vocal tract length is shorter in subjects with DS subject than their controls.

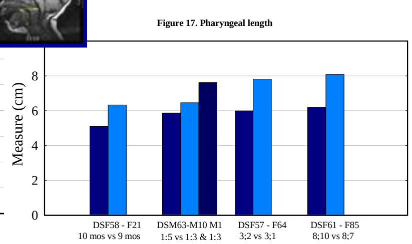
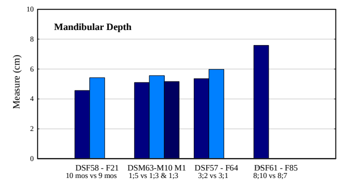
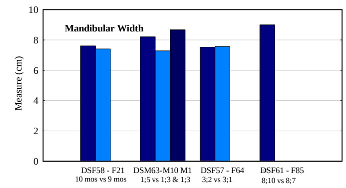
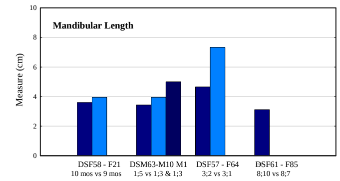


Figure 17. Oro-naso-pharyngeal length is shorter in subjects with DS than their controls.







Figures 18, 19 & 20. Mandibular length and depth are shorter in individuals with DS as compared to their controls. Mandibular width, however, is comparable between the two groups.

Uong et al. 29, who used MRI, report the mandible (mental spine-clivus distance in sagittal plane) to be smaller in DS subjects. This measure captures our mandibular length and depth measures. They report maxillary growth to be reduced in comprison to mandibular growth. Also, they report the measures of maxillary arc (tragion-subnasale-tragion), and upper facial depth (tragion-nasion) to classify 99% DS recognition. Their findings are based on group data from 199 subjects with DS ages 6 months to 61 years.

CONCLUSIONS & DISCUSSION:

- In general, our preliminary findings support other researchers' findings. Imaging technology is adequate to study typical and atypical development of the supralaryngeal speech apparatus.

- To *summarize*, findings indicate:
  Head and face: DS children have a shorter head length, longer head height, shorter upper face height and a flatter facial profile (larger facial convexity measures).
  Vocal tract structures: DS children and their controls have similar tongue length and area, soft palate length, and maxillary length and width. DS children, however, have a shorter hard palate length, shorter mandibular length and depth, shorter oro-nasopharyngeal length, and shorter vocal tract length than their controls.

- These findings, as supported by other researchers, indicate that the many of the physical features/characteristics of the head and neck that are used for diagnostic purposes are based on observations that are not supported by qualitative or metrical research findings. For example, findings do ***NOT*** support the clinical impression of:
  - ***General macroglossia***, but rather relative macroglossia i.e. tongue **appears** bigger because the anterior oral cavity is smaller. Also, since the airway is smaller in DS subjects, the tongue may be positioned forward to open up the airway [17, 29].
  - ***Narrower palate*** -Findings indicate maxillary width to **appear** to be narrower because of its shape. Thus, there is a need to examine the palate in 3D.
  - ***Airway obstruction due to enlarged tonsils*** – instead, findings indicate a smaller airway (due to skeletal abnormalities i.e. smaller midface and lower face skeleton) with smaller adenoid and tonsil volumes [12,29].

- Studies on palatal plate therapy (after Castillo-Morales device) along with speech physiology report long term effects on oral motor function (including inactive open mouth, inactive tongue protrusion) as well as speech improvements [5, 6, 20, 22, 25]. Prospective studies examining the anatomic and physiologic changes that accompany such improvements in speech are indicated. Are improvements due to improved muscle tone of the tongue? Or are there changes in oral anatomy due to the intra-oral forces applied by palatal plate therapy and oral-motor exercises?

- The methodology used to examine anatomic differences between DS subjects and their controls should take into account age. Farkas et al (2002) report that in early childhood (ages 1-5) patients with DS generally displayed more normal than abnormal craniofacial proportions. This corresponds to reports that there are substantial similarities, rather than differences, between the vocalizations of infants with DS and typically developing children [8, 13, 24]. Thus, anatomic, physiologic and acoustic studies that examine the age at which more differences than similarities between DS subjects and their controls emerge are indicated.

Acknowledgments: This work was supported by NIH/NIHCD grant # R03-DC4362 & R01-DC 006282. Special thanks to Cliff Kalina and Gusty Schultz for assistance in scanning the images, and Celia Choih for assistance with figures.

SELECT REFERENCES: Addendum to poster **ASHA November 20, 2004**

MRI Comparison of Children's Vocal Tract Anatomy: Normal vs Down Syndrome - Preliminary data.

By: *Houri K. Vorperian, Ray D. Kent, and Lindell R. Gentry*

1. Allanson, J.E., O'Hara, P., Farkas, L.G. & Nair, R.C. (1993). Anthropometric craniofacial pattern profiles in Down syndrome. *American Journal of Medical Genetics*, 47:748-752.
2. Ardran, G.M., Harker, P. & Kemp, F.H. (1972). Tongue size in Down's syndrome. Journal of Mental Defic Research, 16(84): 160-166.
3. Bleile, K. (1982). Consonant ordering in Down's syndrome pathology. *Journal of Communication Disorders*, 15, 275-285.
4. Borsel, J.V. (1988). An analysis of the speech of five Down's syndrome adolescents. *Journal of Communication Disorders*, 21, 409-421.
5. Carlstedt, K., Dahllof, G., Nilsson, B. & Modeer, T. (1996). Effects of palatal plate therapy in children with Down syndrome. A 1-year study. *Acta Odontologica Scandinavica,* 54(2): 122-125.
6. Carlstedt, K., Henningsson, G. McAllister, A. & Dahllof, G. (2001). Long-term effects of palatal plate therapy on oral motor function in children with Down syndrome evaluated by video registration. *Acta Odontologica Scandinavica,* 59(2): 63-68.
7. Dodd, BJ (1972). Comaprison of babbling patterns in normal and Down-syndrome infants. *Journal of Mental Deficiency Research,* 16, 35-40.
8. Dodd, B. (1976). A comparison of the phonological systems of mental age matched normal, severly subnormal, and Down's syndrome children. *British Journal of Mental Deficiency*, 80, 306-311.
9. Farkas, L.G., Katic, M.J., Forrest, C.R. & Litsas, L. (2001) Anatomical Landmarks: Surface anatomy of the face in Down's syndrome: Linear and angular measurements in craniofacial regions. *Journal of craniofacial surgery* 12(4)373-379
10. Farkas, L. G.; Katic, M. J., and Forrest, C. R. (2001). Surface anatomy of the face in down's syndrome: anthropometric proportion indices in the craniofacial regions. *Journal of Craniofacial Surgery*. 12(6):519-24; discussion 525-6.
11. Farkas, L. G.; Katic, M. J.; Forrest, C. R., and Litsas, L.(2001) Surface anatomy of the face in down's syndrome: linear and angular measurements in the craniofacial regions. *Journal of Craniofacial Surgery*, 12(4):373-9; discussion 380.
12. Jacobs, I.N., Gray, R.F. & Todd, N.W. (1996). Upper airway obstruction in children with Down syndrome. *Archives of Otolaryngology, Head & Neck Surgery*. 122(9) 945-950.
13. Krmpotic-Nemanic, J. and Schulz, J. (1970) Low set ears and small oral cavity with relative macroglossia in down's syndrome: an explanation of the phenomenon. Chicago Medical School Quarterly.; 30(1):41-5.
14. Leddy, M. (1999). The biological bases of speech in people with Down Syndrome. In J.F. Miller, M. Leddy; and L.A. Leavitt (1999). <u>Improving the communication of people with Down Syndrome</u>. Baltimore, Maryland.: Paul H. Brookes Publishing Co., Inc.
15. Miller, J., & Leddy, M. (1998). Down syndrome: The impact of speech production on language development. In R. Paul (Ed.), Communication and language intervention series: Vol. 8 Exploring the speech-language connection (pp. 163-177). Baltimore: Paul H. Brookes Publishing Co.
16. Miolo, G. & Kent, R.D. (1990). Phonological characteristics of babbling in children with Down Syndrome. Presented at 23rd *Annual conference of Research and Theory in Mental Retardation and Developmental Disabilities*, Brainerd, MN.
17. Krmpotic-nemanic, J. & Schulz, J. (1970). Low set ears and small oral cavity with relative macroglossia in Down syndrome: An explanation of the phenomenon. The Chicago Medical School Quarterly, 30(1): 41-45.
18. Mizuno, K. and Ueda, A. (2001) Development of sucking behavior in infants with down's syndrome. *Acta Paediatrica*. 90(12):1384-8.
19. Myrelid, A.; Gustafsson, J.; Ollars, B., and Anneren, G. (2002) Growth charts for down's syndrome from birth to 18 years of age. *Archives of Disease in Childhood*.; 87(2):97-103.
20. Panchon-Ruiz, A., Jornett-Carrillo, V., Sanchez del Campo, F. (2000). Palate vault morphology in Down syndrome. *Journal of Craniofacial Genet Dev Biol*, 20:198-200.
21. Pueschel, S.M. (March 1992) http://TheArc.org/faqs/down.html
22. Pediatric database by A. Gandy (1994) http://www.icondata.com/health/pedbase/files/downsynd.htm
23. Redman, R. S.; Shapiro, B. L., and Gorlin, R. J. (1966) Measurement of normal and reportedly malformed palatal vaults. Ii. Normal juvenile measurements. Journal of Dental Research. 1966 Mar-1966 Apr 30; 45(2):266-9.
24. Rosin, M.M. Swift, E., Bless, D., & Kluppel Vetter, D. (1988). Communication profiles of adolescents with Down syndrome. *Journal of Childhood Disorders*, 12, 49-64.
25. Schuster, G. & Giese, R. (2001). Retrospective clinical investigation of the impact of early treatment of children with Down's syndrome according to Castillo-Morlaes. *Journal of Orofacial Orthopedics*, 62(4): 255-263.
26. Shapiro, B. L. (1975). Amplified developmental instability in Down's syndrome. *Annals of Human Genetics*. 38(4): 429-437.
27. Shapiro, B. L.; Gorlin, R. J.; Redman, R. S., and Bruhl, H. H. (1967) The palate and Down's syndrome. *New England Journal of Medicine*.; 276(26): 1460-3.
28. Smith, B.L. & Oller, D.K. (1981). A comparative study of premeaningful vocalizations produced by normally developing and Down's syndrome infant. *Journal of Speech and Hearing Disorders*, 46, 46-51.
29. Uong, E.C., McDonough, J.M., Tayag-Kier, C.E., Zhao, H., Haselgrove, J. Mahboubi, S., Schwab, R.J. Pack, A.I. & Arens, R. (2001). Magnetic resonance imaging of the upper airway in children with Down Syndrome. *American Journal of Respir Crit Care Med* 163: 731-736.
30. Vorperian, H.K., Kent, R.D., Gentry, L.R. & Yandell, B.S. (1999). MRI procedures to study the concurrent anatomic development of the vocal tract structures: Preliminary results. *International Journal of Pediatric Otorhinolaryngology*, 49(3) 197-206.