Research Article

Speech, Language, and Communication Skills of Adolescents With Craniofacial Microsomia

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Purpose: Youth with craniofacial microsomia (CFM) have anomalies and comorbidities that increase their risk for speech, language, and communication deficits. We examined these outcomes in youth with and without CFM and explored differences as a function of CFM phenotype and hearing status.

Method: Participants included youth ages 11–17 years with CFM ($n = 107$) and demographically similar controls ($n = 306$). We assessed speech intelligibility, articulation, receptive and expressive language, and parent and teacher report measures of communication. Hearing status was also screened at the study visit. Group differences were estimated using linear regression analyses with standardized effect sizes (ES) adjusted for demographic characteristics (adjusted ES) or negative binomial regression.

Results: Youth with CFM scored lower than unaffected peers on most measures of intelligibility, articulation, expressive language, and parent- and teacher-rated communication. Differences were most pronounced among participants with CFM who had mandibular hypoplasia plus microtia (adjusted ES = -1.15 to -0.18). Group differences were larger in youth with CFM who failed the hearing screen (adjusted ES = -0.73 to 0.07) than in those who passed the hearing screen (adjusted ES = -0.34 to 0.27).

Conclusions: Youth with CFM, particularly those with mandibular hypoplasia plus microtia and/or hearing loss, should be closely monitored for speech and language concerns. Further research is needed to identify the specific needs of youth with CFM as well as to document the course of speech and language development in children with CFM.

Craniofacial microsomia (CFM), also known as hemifacial microsomia, is a congenital condition characterized by asymmetric underdevelopment of the mandible and ear, although other cranial and extra-cranial malformations may also be observed (e.g., cleft palate, vertebral anomalies). CFM ranges in severity from mild to severe facial asymmetry involving multiple structures. CFM affects approximately one in 3,500–5,600 live births (Barisic et al., 2014), with higher prevalence among boys

and children of Hispanic and Native American descent (Harris, Kallen, & Robert, 1996). Individuals with CFM often have complex health care needs and are best served by a multidisciplinary craniofacial team. In early childhood, patients with CFM require audiologic monitoring and intervention for hearing loss (e.g., management of Eustachian tube dysfunction, hearing aids), optimization of feeding and growth, and evaluation for symptoms of upper airway obstruction. Treatments in middle childhood through late adolescence often include ear and mandibular reconstruction. Throughout development, monitoring children with CFM for speech and language concerns, psychological adjustment, and educational progress is recommended (American Cleft Palate Craniofacial Association, 2018).

Previous studies have shown neurodevelopmental differences in children with CFM relative to test norms and unaffected peers (M. S. Cohen et al., 1995; Collett et al., 2011; Speltz et al., 2017). In our own research, we found that school-aged children with CFM scored significantly lower on teacher-administered neurocognitive screening measures than unaffected children (Collett et al., 2011). Differences were largest on the measure of receptive vocabulary,

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with children with CFM almost five times more likely to score in the “delayed” range than children without CFM. Neurocognitive differences were most apparent in children with reported speech, hearing, and/or vision impairments.

Several factors may compromise speech and language development for children with CFM. Most individuals with CFM (~75%–80%) have some degree of hearing impairment, usually in the form of conductive hearing loss, although sensorineural hearing loss has been observed in approximately 10% of children with CFM (Mitchell et al., 2017; Rahbar et al., 2001; Vrabec & Lin, 2010; Wan, Meara, Kovanlikaya, Nelson, & Don, 2003). Although the associations between hearing impairment and speech and language development have not been well-studied in CFM, there is extensive literature on the general population showing that even mild unilateral hearing loss is associated with higher prevalence of delays in speech articulation and receptive and expressive language delay, learning disabilities, and poorer communication and social skills (Anne, Lieu, & Cohen, 2017; Lieu, 2018). Children with CFM may incur additional risk of articulation errors, independent of hearing loss, as a result of one or more associated craniofacial anomalies including facial nerve palsy, asymmetric soft palate movement, orofacial clefting, malocclusion, tongue anomalies, and/or muscular deficiencies (Carvalho, Song, Vargervik, & Lalwani, 1999; Chen et al., 2009; Shprintzen, Croft, Berkman, & Rakoff, 1980; Strömberg et al., 2007). These structural factors may impact children’s production of speech sounds, overall speech intelligibility/acceptability, vocal quality, and/or resonance. Finally, stigma associated with visible facial differences and differences in articulation may interfere with children’s social engagement and communication with peers, teachers, and other adults (Bhaskute et al., 2017; Bickham et al., 2017).

Despite these anticipated vulnerabilities, we are not aware of any previous studies of speech and language development in individuals with CFM that included a comparison group of unaffected peers. We therefore sought to compare speech, language, and communication outcomes of adolescents with CFM to a sample of demographically similar peers without CFM. This study followed into adolescence, the same cohort of children with and without CFM tested by Collett et al. (2011). We used a multimethod assessment approach, including standardized testing of articulation and language, administered by trained psychometrists, and parent and teacher reports of communication skills at home and school. We addressed two primary questions: (a) In comparison with unaffected peers, do adolescents with CFM demonstrate impaired articulation, language, and communication skills? (b) Do phenotypic factors (e.g., anatomic variations, hearing status) contribute to differences in communication between youth with CFM and unaffected peers?

Method

Study Design

The Child and Adolescent Learning and Living Study is Phase 3 of an observational investigation of prenatal risk

factors and neurodevelopment in youth with and without CFM (Werler et al., 2004). Specific group participation and attrition numbers for all three phases of this research and reasons for participant loss have previously been published (Speltz et al., 2017). Participants < 48 months of age were enrolled between 1996 and 2002 from 26 cities across the United States and Canada. Child and Adolescent Learning and Living Study Phase 1 was a case-control study to identify demographic and risk factors for CFM (Werler et al., 2004). A longitudinal component was added in Phase 2, which involved neurodevelopmental and psychosocial assessments when the children were on average 7 years of age (Collett et al., 2011). These participants were reapproached for the current (Phase 3) study between 2011 and 2015, when youth were 13 years of age on average (range: 11–17 years). Phase 3 featured a comprehensive assessment of multiple neuropsychological, educational, and other outcomes; the present report describes a subset of these measures that are related to articulation, language, and communication. All participating youth were fluent in English, although Spanish was the preferred language for 26 parents. The study was approved by the institutional review boards of both participating centers (Seattle Children’s Hospital and Boston University).

Youth With CFM

In Phase 1, children with CFM were ascertained from craniofacial clinics and were eligible if they were less than age 48 months and had received a diagnosis of CFM, facial asymmetry, unilateral microtia, oculo-auriculo-vertebral spectrum, or Goldenhar syndrome from a craniofacial physician. We excluded children with diagnosed syndromes (e.g., Treacher Collins syndrome) and those with known chromosomal anomalies. Participants without microtia and/or at least two CFM-associated malformations were excluded ($n = 21$) from the current analyses in order to include a more homogeneous sample of youth with the most prototypical features of CFM.

Unaffected Control Peers

Families of infants without CFM (“controls”) were recruited through the pediatricians’ offices of participants with CFM or from another pediatric practice in proximity. Approximately, three control families were recruited for each participant with CFM in Phase 1. Controls were eligible if they had no known birth defects, had not been adopted, and were within 2 months of the matched affected subject’s age at the time of recruitment. For Phase 3, we selected two or three matching group participants for each participant with CFM depending on geographic proximity. When there were several potential eligible unaffected peers in a given location, we prioritized controls that were most similar to the target child with CFM in age, sex, and language spoken in the home (English or Spanish).

Participants included 121 youth with CFM and 315 youth without CFM, representing 72% and 64% of those approached for participation. Compared to participants, nonparticipants were more likely to be non-White

(23% participants, 37% nonparticipants) and less likely to be from predominately Spanish-speaking households (6% participants, 10% nonparticipants). Nonparticipants were more likely than participants to have scored in the “delayed” range on one or more neurodevelopmental measures during the previous phase of this research (18% participants, 28% nonparticipants). Articulation and language data were missing for some participants (14 youth with CFM, nine controls) due to missing or unusable video recordings, participant or family time constraints that precluded the administration of some tests, or severe developmental delay that limited test participation. In total, data were available for one or more outcomes for 107 youth with CFM and 306 controls.

Assessment Procedures

Participants were assessed by trained psychometrists in community settings (e.g., libraries, schools, community centers) or in their homes. Testing sessions lasted approximately 3 hr. All testing was recorded using a digital video recorder (Canon Vivia HFM 30) and a desktop microphone (Shure Microflex 412D/C).

Measures

Language and Communication

Expressive and receptive language skills were assessed using three core language subtests from the Clinical Evaluation of Language Fundamentals–Fourth Edition (CELF-4; Semel, Wiig, & Secord, 2003): (a) Recalling Sentences, which assesses the ability to verbally recall sentences of variable length and complexity; (b) Word Classes 2–Receptive, which assesses the ability to identify relationships between orally presented target words; and (3) Word Classes 2–Expressive, which measures a child’s ability to explain the association between target words. Norm-referenced scaled scores were calculated for each subtest (M scaled score = 10, SD = 3) and used in the statistical analyses. A randomly selected 20% of the CELF-4 administrations were reviewed for reliability by one of the authors (B. C.), including listening to the recordings and rescored the CELF-4 subtests. Item-level agreement was $\geq 98\%$ for all subtests.

Parents and teachers completed the Children’s Communication Checklist–Second Edition (CCC-2; Bishop, 2003), a norm-referenced measure of communication skills. We compared scores for youth with and without CFM for speech and language subtests (Speech, Syntax, Semantics, Coherence, Nonverbal Communication), as well as the General Communication Composite (GCC). Subtests use scaled scores (M = 10, SD = 3), whereas the GCC uses a standard score (M = 100, SD = 15). Outcomes for one subtest, Social Relations and Interests, were reported separately in a paper on social and behavioral outcomes (Wallace, Collett, Heike, Werler, & Speltz, 2018). A Spanish version of the CCC-2 is not available, and this measure was therefore not administered to participants whose parents spoke primarily Spanish.

Speech Intelligibility and Articulation

Speech intelligibility was assessed using the Speech Intelligibility Test (SIT), a computer-based measure of intelligibility, speaking rate, and communication efficiency calculated from samples of spoken sentences (Yorkston, Beukelman, Hakel, & Dorsey, 2007). Participants were presented a list of 11 unique sentences (randomly chosen from a database of 1,100 different sentences) to repeat after the examiner. Participants’ productions of novel sentences were video-recorded and later played back (via headphones) to a master’s level speech-language pathologist (SLP). Although speech samples were recorded on video, only audio was used and the SLP was unaware of participants’ CFM status. The SLP typed out the sentences they heard and compared the transcriptions against the target sentence. A percent intelligibility score was then calculated by the SIT computer program. When there was not a 100% match between the listeners’ transcription and the computer-generated sentence, an examiner manually checked the disagreements. Spelling or other transcription errors were excluded from percent intelligibility scores.

Forty-five sentences from the Templin–Darley Tests of Articulation (TDTA; Templin & Darley, 1969) sentences were employed to assess articulation. Sentences were chosen to assess all singleton (across all word positions) and common consonant clusters present in English. Sentences were then phonetically transcribed using the International Phonetic Alphabet (International Phonetic Association, 2001) by master’s level SLPs. Errors related to reading difficulty were not included in the counts of speech production errors. Percentage of correct productions was calculated for each participant’s sample. Perceptual ratings of voice and resonance were also made by a licensed SLP using the TDTA sentence sample. Adolescents’ speech was rated for degree of hypernasality on an equal-appearing interval 4-point scale (0 = *absent*, 1 = *minimal/mild hypernasality*, 2 = *moderate hypernasality*, 3 = *severe hypernasality*). Hyponasality was rated as present or absent. Finally, vocal quality was also rated as normal or atypical.

A randomly selected 20% of the samples were independently transcribed to calculate interrater reliability. Percentage agreement scores were calculated for phoneme match on the TDTA, for word match judgments on the SIT, and for judgments of resonance and vocal quality. Similarly, a randomly selected 20% of the samples were scored for intrarater reliability, with the same SLP scoring the measure twice approximately one month apart. Mean inter- and intrarater reliability scores were 99% for both measures on the TDTA. Word matches on the SIT were 96% and 97% for inter- and intrarater reliability, respectively. Agreement scores for resonance ratings were 100% and 99% for inter- and intrarater agreement, respectively. Finally, interrater reliability for vocal quality was 98% and intrarater reliability was 99%.

Audiometric Assessment

Conventional pure-tone screening audiometry was used to screen for hearing loss among participants with

and without CFM using a GSI 18 portable device (American Speech-Language-Hearing Association, 2015). Trial presentations of tones at 45–55 dB HL were used to confirm participants' understanding of the procedure. Both ears were tested at 500, 1000, 2000, and 4000 Hz. The participant's performance was considered a "pass" if he or she responded at ≤ 40 dB HL for any of these frequencies in both ears. These criteria were chosen as a hearing level of ≤ 40 dB HL and were used to classify those individuals with no more than mild hearing loss from those with moderate or more severe hearing impairment (American Speech-Language-Hearing Association, 2015).

Phenotypic Assessment

A detailed description of our methodology for phenotypic assessments, as well as subgroup identification for this study cohort, is described elsewhere (Birgfeld et al., 2016; Heike et al., 2016). Briefly, phenotypic classifications were developed based on the Orbital, Mandible, Ear, Nerve, Soft Tissue (OMENS) pictorial rating scale (Poon, Meara, & Heggie, 2003; Tuin et al., 2015). The OMENS scale groups cases were based on morphological features of CFM with shared clinical implications. To complete these ratings in our geographically diverse epidemiological sample, we collected standardized facial photographs of all participants at Phase 3. This protocol consisted of 16 views of the face and a classification method described by Birgfeld et al. (2016). Ratings of photos using this method have correlated highly with physical examination for most features, and we obtained interrater reliability kappa coefficients of .7 or greater for each of the OMENS features using physician raters (Birgfeld et al., 2013). In the current study, a craniofacial pediatrician rated all photographs. Additionally, we integrated data from medical chart information obtained during Phase 1 and data from a medical questionnaire obtained at the Phase 3 study visit. For each feature on each study participant, data from all three sources (i.e., photographic ratings, medical questionnaire, medical chart abstraction) were reviewed in order to establish phenotype. Three mutually exclusive phenotypic subgroups were identified a priori: (a) microtia only (i.e., absence of mandibular hypoplasia, epibulbar dermoids, lateral clefts, preauricular or facial tags, small and/or displaced orbit, nerve palsies), (b) microtia and mandibular hypoplasia, and (c) other combinations of CFM-associated malformations (≥ 2 malformations were required).

Medical History Interview and Questionnaire

During Phase 1, mothers of youth with CFM were interviewed by telephone about demographic, reproductive, and medical factors. At Phase 3, mothers or guardians completed a questionnaire to update the family's demographic information and medical history (including patients' medical/surgical histories) and to provide information on any developmental or educational interventions or accommodations received (e.g., speech-language, audiology, vision, physical or occupational therapy, or other special education services).

Statistical Analyses

We calculated descriptive statistics (means, standard deviations, frequencies) separately for youth with and without CFM to summarize demographic characteristics and scores for all outcomes. To estimate overall group differences and corresponding 95% confidence intervals, we used linear regression with robust standard errors for all continuous outcomes with the exception of percent consonant correct on the TDTA sentences. We estimated the magnitude of adjusted standardized group differences (standardized effect size [ES]) by calculating a modification of Cohen's *d*, using the adjusted mean difference and dividing by the root-mean-square error for the model (J. Cohen, 1988). Negative binomial regression was used to estimate group differences in the number of TDTA errors to account for data overdispersion. Covariates, selected a priori, included youths' age at assessment (in years), sex and race/ethnicity (non-Hispanic White, Hispanic, and other), and family income and the primary caregiver's highest level of education. In separate linear regression analyses, we examined group differences as a function of CFM phenotype (microtia only, microtia plus mandibular hypoplasia, other CFM-related anomalies) and an indicator for a failed hearing screening. Controls served as the referent category for all of these analyses and covariates included the variables listed above. In addition to examining differences for each phenotypic or hearing status group, we used Wald tests to determine overall group differences.

In sensitivity analyses, we examined the effect of attrition bias by repeating the primary analysis using inverse probability weighting (Robins, Rotnitzky, & Zhao, 1994). The predicted probability of participation in Phase 3 (vs. nonparticipation due to refusal or loss of contact) was estimated with characteristics collected at the school-age assessment or earlier including CFM status, gender, race, language spoken at home (Spanish vs. English), and data from neurodevelopmental tests and behavior questionnaires. We also conducted sensitivity analyses that excluded all participants from predominantly Spanish-speaking homes. Finally, we used censored normal regression to examine the potential effects of speech and language interventions on observed group differences (Tobin, Sheehan, Scurrah, & Burton, 2005). This approach assumes that the scores of youth who received interventions would be at least as low as those observed in the absence of intervention, that is, that they are "left-censored."

Results

Youth with CFM were more likely than controls to be male and Hispanic, and caregivers' highest level of education and family income were both slightly lower among families of youth with CFM versus controls (see Table 1). Relative to controls, youth with CFM were more likely to have received speech therapy (60% of youth with CFM, 16% of controls) and/or to have ever used hearing aids (23% of youth with CFM, 1% of controls). Seventy percent of

Table 1. Characteristics of adolescents with and without CFM.

Characteristic	Controls (<i>N</i> = 315)		Cases (<i>N</i> = 121)	
	<i>N</i>	%	<i>N</i>	% ^a
Age in years				
< 12	64	20.4	19	16.0
12–14	152	48.4	59	49.6
> 14	98	31.2	41	34.5
Gender				
Female	162	51.4	44	36.4
Male	153	48.6	77	63.6
Race/ethnicity				
White	251	79.7	87	71.9
Hispanic	33	10.5	28	23.1
Black	19	6.0	0	0.0
Asian/Pacific Islander	9	2.9	4	3.3
Native American	3	1.0	2	1.7
Grade				
5–6	101	33.7	42	35.9
7–8	127	42.3	55	47.0
9–12	70	23.3	20	17.1
Family income				
< \$25,000	32	10.2	17	14.4
\$25,000–\$34,999	23	7.3	20	16.9
\$35,000–\$64,999	47	15.0	19	16.1
≥ \$65,000	211	67.4	62	52.5
Primary caregiver's highest level of education				
< 12 years	11	3.5	9	7.7
High school/GED	73	23.3	43	36.8
Associate's degree	59	18.8	17	14.5
Bachelor's degree	104	33.2	37	31.6
Graduate degree	66	21.1	11	9.4
Site				
Boston	221	70.2	74	61.2
Seattle	94	29.8	47	38.8
Phenotype case group				
Microtia only	0	0.0	24	19.8
Microtia and mandibular hypoplasia	0	0.0	46	38.0
Other CFM-associated anomalies	0	0.0	51	42.1
No discernible anomaly	315	100.0	0	0.0
Isolated cleft palate or cleft lip plus cleft palate	0	0.0	19	15.7
Hearing impairment ^b	2	0.7	77	70.0
Ever used hearing aids ^c	2	0.6	28	23.0
Receipt of speech/language interventions	47	15.8	68	59.6

Note. CFM = craniofacial microsomia; GED = General Educational Development.

^aPercentages represent proportion with nonmissing values. Missing data: age (one control, two cases), grade (15 controls, four cases), income (two controls, three cases), education (two controls, four cases), hearing impairment status (15 controls, 11 cases), ever used hearing aids (seven controls, one case), and speech and language intervention (18 controls, seven cases). ^bDefined as a hearing threshold > 40 dB at frequencies of 500, 1000, 2000, or 4000 Hz; note values coded as missing due to room noise or missing test. ^cHistory of hearing aid use was self-reported and only asked in families who reported a history of hearing loss.

youth with CFM failed the hearing screen compared to 1% of controls. Among participants with CFM, 20% had microtia only, 38% had microtia plus mandibular hypoplasia, and 42% had other CFM-related facial anomalies. Sixteen percent of youth with CFM had an orofacial cleft.

Group Differences

Articulation and intelligibility. Articulation errors were infrequent in both groups (see Table 2). On the TDTA, youth with CFM produced 94% of target sounds correctly compared to 99% correct among controls (adjusted ES =

−0.98, $p < .001$; see Table 3). Descriptively, participants with CFM were more likely than those without CFM to exhibit disordered speech (35% vs. 3%), hypernasality (14% vs. 1%), hyponasality (4% vs. 0%), and disordered vocal quality (7% vs. 1.4%). Intelligibility was high on the SIT in both groups (93% correct for youth with CFM, 97% correct for youth without CFM), though significantly lower in participants with CFM compared to unaffected peers (adjusted ES = −0.61, $p = .001$).

Receptive and expressive language. Participants with CFM scored slightly lower than controls on each of the CELF-4 subtests, though differences were small in magnitude

Table 2. Distribution of TDTA and SIT outcomes in adolescents with and without craniofacial microsomia.

Test and measure	Controls		% or SD	Cases		% or SD
	Total N	N or M		Total N	N or M	
TDTA						
Percent correct	286	99.17	2.14	95	93.67	9.79
Speech category						
Normal	288	279	96.9	99	64	64.6
Disordered	288	9	3.1	99	35	35.4
Resonance category						
Normal	288	285	99.0	96	83	86.5
Mild hypernasality	288	3	1.0	96	11	11.5
Moderate hypernasality	288	0	0.0	96	1	1.0
Severe hypernasality	288	0	0.0	96	1	1.0
Hyponasality	288	0	0.0	96	4	4.2
Voice category						
Normal	288	284	98.6	100	93	93.0
Disordered	288	4	1.4	100	7	7.0
SIT						
Percent correct	283	97.4	2.9	97	92.6	12.4

Note. TDTA = Templin–Darley Tests of Articulation; SIT = Speech Intelligibility Test.

and the only statistically significant difference was on Recalling Sentences (adjusted ES = -0.27 , $p = .02$; see Table 3).

Parent and teacher communication ratings. There were no differences in parent ratings for any of the language subtests of the CCC-2 or the GCC (adjusted ES = -0.17 to 0.04 , $p = .20$ – $.74$). However, parent ratings indicated that

youth with CFM had lower scores than controls on the CCC-2 Speech subtest (adjusted ES = -0.39 , $p = .004$; see Table 3). Teachers rated youth with CFM lower than controls on the Speech (adjusted ES = -0.45 , $p = .003$), Semantics (adjusted ES = -0.29 , $p = .03$), and Nonverbal Communication subtests (adjusted ES = -0.39 , $p = .01$; see Table 3).

Table 3. Comparison of mean speech and language scores for adolescents with and without craniofacial microsomia.

Test and measure	Unadjusted		Adjusted ^c				
	Mean diff.	ES ^d	Mean diff.	95% CI	ES ^d	p value	
CELF (306 controls, 107 cases) ^{a,b}							
Recalling Sentences	−1.38	−0.46	−0.76	−1.41 −0.10	−0.27	.02	
Receptive	−1.20	−0.41	−0.56	−1.20 0.08	−0.21	.09	
Expressive	−1.13	−0.36	−0.41	−1.06 0.24	−0.14	.22	
CCC2-Parent (288 controls, 96 cases) ^a							
Speech	−1.30	−0.45	−1.09	−1.82 −0.35	−0.39	.004	
Syntax	−0.41	−0.17	−0.16	−0.72 0.41	−0.07	.59	
Semantics	−0.16	−0.06	0.09	−0.46 0.65	0.04	.74	
Coherence	−0.48	−0.18	−0.26	−0.89 0.38	−0.10	.43	
Nonverbal Communication	−0.60	−0.25	−0.38	−0.95 0.20	−0.17	.20	
General Communication Composite	−3.16	−0.20	−1.51	−5.05 2.03	−0.10	.40	
CCC2-Teacher (237 controls, 80 cases) ^a							
Speech	−1.58	−0.54	−1.26	−2.07 −0.45	−0.45	.003	
Syntax	−0.77	−0.30	−0.52	−1.18 0.15	−0.21	.13	
Semantics	−1.02	−0.40	−0.71	−1.35 −0.07	−0.29	.03	
Coherence	−0.56	−0.21	−0.25	−0.95 0.46	−0.10	.49	
Nonverbal Communication	−1.28	−0.50	−0.97	−1.70 −0.24	−0.39	.01	
General Communication Composite	−5.92	−0.40	−3.84	−7.73 0.06	−0.27	.05	
TDTA (286 controls, 95 cases) ^b							
Percent correct	−5.50	−0.105	−5.07	−7.08 −3.06	−0.98	< .001	
SIT (283 controls, 97 cases)							
Percent correct	−4.79	0.71	−4.05	−6.36 −1.73	−0.61	.001	

Note. diff = difference; CI = confidence interval; CELF = Clinical Evaluation of Language Fundamentals; CCC-2 = Children's Communication Checklist-2; TDTA = Templin–Darley Tests of Articulation; SIT = Speech Intelligibility Test.

^aScaled scores used for all CELF and CCC-2 measures, with the exception of the CCC-2 General Communication Composite, which uses standard scores ($M = 100$, $SD = 15$). ^bOne control was missing CELF Receptive, Expressive, and TDTA percent correct data. ^cAdjusted for age (continuous), gender, race (White/non-White), income (categorical), and primary caregiver's highest level of education (categorical). ^dES = standardized effect size.

Group Differences as a Function of CFM Phenotype and Hearing Status

Participants with CFM who had microtia plus mandibular hypoplasia scored lower than control peers across all measures (see Table 4). Deficits in this group were moderate to large in magnitude for the SIT, CELF-4 Recalling Sentences, and CCC-2 Speech and Nonverbal Communication subtests for both parent and teacher reports, as well as the Semantics and GCC subtests reported by teachers ($ES = -1.15$ to -0.53 , $p < .001-.01$). Although ES were modest, and not all differences were statistically significant, youth with CFM who had microtia plus mandibular hypoplasia also scored lower than controls on all other outcome measures ($ES = -0.48$ to -0.18 , $p = .01-.32$). The CELF-4, CCC-2 and SIT scores were similar between youth with CFM who had microtia only relative to controls ($ES = -0.33$ to 0.30 , $p = .17-.68$), as were scores for youth with CFM characterized by other CFM-associated features relative to controls ($ES = -0.26$ to 0.21 , $p = .17-.98$). TDTA scores were higher across all phenotype groups compared to controls, on average by 1.5 to 2 points (all $p < .001$). Among the three phenotypes, youth with CFM characterized by microtia plus mandibular hypoplasia exhibited the highest proportion of disordered speech (51%), followed by youth with microtia only (30%) and other CFM-associated features (24%).

Seventy percent of participants with CFM did not pass the hearing screening, indicating that they had at least mild hearing loss. The prevalence of hearing impairment was highest in cases with microtia plus mandibular hypoplasia (84%), followed by microtia only (76%) and other CFM-associated features (54%). Group differences on all outcomes were larger for participants with CFM, who failed the hearing screening compared to those who passed. The greatest differences were observed for the SIT (adjusted $ES = -0.73$, $p = .005$), the Speech subtests from the CCC-2 (adjusted $ES = -0.57$ to -0.36 , $p = .002-.03$), and the Nonverbal Communication subtest from the teacher CCC-2 (adjusted $ES = -0.39$, $p = .04$; see Table 5). With the exception of the TDTA, scores for youth with CFM who passed the hearing screening were similar to controls across all other measures, and confidence intervals included the null.

Sensitivity Analyses

After accounting for the effects of attrition using inverse probability weighting, group differences increased in magnitude across all CELF-4 subtests and the SIT ($ES = -0.77$ to -0.30 , $p < .001-.03$; see Table 6). Estimates across other measures were similar to primary analyses. Differences between youth with and without CFM across teacher CCC-2 subtests, with the exception of the GCC,

Table 4. Comparison of mean speech and language scores for adolescents with and without CFM, by phenotype.

Test and measure	Microtia/anotia only					Microtia + mandibular hypoplasia					Other CFM-associated anomalies				
	Mean diff. ^b	95% CI	ES ^c	p value		Mean diff. ^b	95% CI	ES ^c	p value		Mean diff. ^b	95% CI	ES ^c	p value	
CELF ^a															
Recalling Sentences	-0.37	-1.63	0.90	-0.14	.57	-1.79	-2.82	-0.76	-0.65	.001	0.01	-0.86	0.88	0.00	.98
Receptive	-0.17	-1.30	0.96	-0.06	.77	-1.06	-2.16	0.03	-0.40	.06	-0.28	-1.13	0.58	-0.10	.53
Expressive	-0.40	-1.45	0.65	-0.14	.46	-0.97	-2.11	0.17	-0.35	.09	0.11	-0.74	0.96	0.04	.80
CCC2-Parent ^a															
Speech	0.44	-0.58	1.47	0.16	.40	-2.38	-3.51	-1.24	-0.87	< .001	-0.37	-1.31	0.57	-0.13	.44
Syntax	0.19	-0.94	1.31	0.09	.74	-0.74	-1.62	0.14	-0.34	.10	0.29	-0.42	1.00	0.13	.43
Semantics	0.34	-0.96	1.64	0.14	.61	-0.42	-1.26	0.41	-0.18	.32	0.51	-0.17	1.19	0.21	.14
Coherence	0.46	-0.63	1.56	0.18	.41	-1.21	-2.24	-0.19	-0.47	.02	0.42	-0.29	1.13	0.16	.25
Nonverbal Comm.	0.43	-0.54	1.40	0.19	.39	-1.19	-2.10	-0.28	-0.53	.01	0.13	-0.62	0.87	0.06	.74
General Comm.	4.26	-1.75	10.26	0.30	.17	-6.77	-12.15	-1.39	-0.48	.01	1.57	-2.62	5.77	0.11	.46
CCC2-Teacher ^a															
Speech	0.42	-0.93	1.77	0.15	.54	-2.56	-3.85	-1.27	-0.92	< .001	-0.80	-1.91	0.31	-0.29	.16
Syntax	0.26	-1.07	1.59	0.10	.71	-1.13	-2.16	-0.11	-0.46	.03	-0.29	-1.23	0.65	-0.12	.55
Semantics	0.37	-0.80	1.53	0.15	.54	-1.48	-2.48	-0.48	-0.61	.004	-0.48	-1.30	0.34	-0.20	.26
Coherence	0.54	-0.91	1.98	0.21	.47	-1.22	-2.21	-0.23	-0.48	.02	0.31	-0.66	1.28	0.12	.53
Nonverbal Comm.	0.31	-1.17	1.80	0.13	.68	-2.05	-3.06	-1.03	-0.83	< .001	-0.56	-1.60	0.48	-0.23	.29
General Comm.	2.51	-5.61	10.63	0.18	.55	-10.08	-15.79	-4.36	-0.72	.001	-0.87	-5.90	4.15	-0.06	.73
TDTA															
Percent correct	-4.33	-8.12	-0.53	-0.85	.03	-7.70	-11.31	-4.09	-1.51	< .001	-2.98	-5.64	-0.32	-0.58	.03
SIT															
Percent correct	-2.16	-6.55	2.24	-0.33	.34	-7.47	-12.48	-2.47	-1.15	.004	-1.66	-4.06	0.73	-0.26	.17

Note. CFM = craniofacial microsomia; diff. = difference; CI = confidence interval; CELF = Clinical Evaluation of Language Fundamentals; CCC-2 = Children's Communication Checklist-2; Comm. = Communication; TDTA = Templin-Darley Tests of Articulation; SIT = Speech Intelligibility Test.

^aScaled scores used for all CELF and CCC-2 measures, with the exception of the CCC-2 General Communication Composite, which uses standard scores ($M = 100$, $SD = 15$). ^bAdjusted for age (continuous), gender, race (White/non-White), income (categorical), and primary caregiver's highest level of education (categorical). ^cES = standardized effect size.

Table 5. Comparison of mean test scores for adolescents with and without craniofacial microsomia, by hearing impairment.

Test and measure	Hearing impairment (hearing screen > 40 dB) ^a							
	No (N = 33)				Yes (N = 77)			
	Mean diff. ^c	95% CI	ES ^d	p value	Mean diff. ^c	95% CI	ES ^d	p value
CELF ^b								
Recalling Sentences	-0.56	-1.62 0.50	-0.20	.30	-0.77	-1.58 0.04	-0.28	.06
Receptive	-0.16	-1.09 0.76	-0.06	.73	-0.60	-1.40 0.21	-0.23	.15
Expressive	0.02	-0.91 0.95	0.01	.97	-0.45	-1.27 0.37	-0.16	.28
CCC2-Parent ^b								
Speech	-0.97	-2.21 0.26	-0.34	.12	-1.02	-1.93 -0.11	-0.36	.03
Syntax	-0.10	-0.90 0.70	-0.05	.80	0.02	-0.70 0.75	0.01	.95
Semantics	0.13	-0.73 1.00	0.05	.77	0.17	-0.52 0.86	0.07	.64
Coherence	0.18	-0.82 1.17	0.07	.73	-0.43	-1.21 0.35	-0.16	.28
Nonverbal Comm	0.14	-0.68 0.95	0.06	.74	-0.55	-1.31 0.20	-0.24	.15
General Comm.	-0.17	-5.66 5.32	-0.01	.95	-1.44	-5.75 2.87	-0.10	.51
CCC2-Teacher ^b								
Speech	-0.79	-2.08 0.51	-0.28	.24	-1.60	-2.62 -0.58	-0.57	.002
Syntax	-0.41	-1.49 0.67	-0.17	.46	-0.40	-1.18 0.39	-0.16	.32
Semantics	-0.50	-1.57 0.57	-0.20	.36	-0.74	-1.53 0.04	-0.30	.06
Coherence	0.70	-0.41 1.81	0.27	.22	-0.60	-1.48 0.28	-0.23	.18
Nonverbal Comm.	-0.47	-1.62 0.68	-0.19	.43	-0.97	-1.87 -0.07	-0.39	.04
General Comm.	-0.98	-7.30 5.34	-0.07	.76	-4.62	-9.41 0.18	-0.33	.06
TDTA								
Percent correct	-3.64	-6.89 -0.40	-0.72	.03	-5.40	-7.84 -2.96	-1.07	< .001
SIT								
Percent correct	-1.74	-3.59 0.11	-0.27	.07	-4.69	-7.92 -1.45	-0.73	.005

Note. diff. = difference; CI = confidence interval; CELF = Clinical Evaluation of Language Fundamentals; CCC-2 = Children's Communication Checklist-2; Comm. = Communication; TDTA = Templin-Darley Tests of Articulation; SIT = Speech Intelligibility Test.

^aTwo controls with hearing impairment excluded from analysis; 12 controls and 10 cases missing hearing screen; three controls and one case excluded due to room noise. ^bScaled scores used for all CELF and CCC-2 measures, with the exception of the CCC-2 General Communication Composite, which uses standard scores ($M = 100$, $SD = 15$). ^cAdjusted for age (continuous), gender, race (White/non-White), income (categorical), and primary caregiver's highest level of education (categorical). ^dES = standardized effect size.

increased slightly after excluding Spanish-speaking participants ($ES = -0.60$ to -0.19 , $p < .001-.18$), but there was little shift in ES for the CELF-4, TDTA, or SIT outcomes (see Table 6). After accounting for the effects of interim speech and language therapy using censored normal regression, group differences increased in magnitude across all measures (see Table 6).

Discussion

To our knowledge, this is the first large study of speech, language, and communication outcomes in a cohort of youth with and without CFM. Participants were assessed in early adolescence, an age when normal speech is close to 100% intelligible and nearly all speech sounds are correctly produced (Smit, Hand, Freiling, Bernthal, & Bird, 1990). In order to characterize participants' morphological features and to better understand potential associations between facial structure and speech functions, we used a detailed phenotypic assessment method based on multiple data sources (Heike et al., 2016). Our findings suggest that youth with CFM exhibit lower speech intelligibility and more disordered articulation than unaffected peers, as well as modest impairments in receptive language and communication as rated by parents and teachers. Group differences were similar or slightly larger in analyses excluding children from

predominately Spanish-speaking homes. Results of sensitivity analyses to account for interim intervention services and differential attrition suggest that our findings may underestimate the true population differences between youth with CFM and their unaffected peers. That is, observed differences would be expected to be larger in higher social risk populations (e.g., among predominantly low socioeconomic status youth) and in youth with limited access to speech and language interventions.

The underlying reasons for these differences are unclear, though our assessments of phenotypic subgroups and outcomes as a function of hearing status provide some clues and directions for future research. Group differences were limited to youth with CFM who have both microtia plus mandibular hypoplasia; youth with microtia only and those with other CFM-associated anomalies were similar to controls. Thus, characteristics related to mandibular hypoplasia (e.g., limited range of motion, asymmetry of mandibular movements) may limit the ability to establish accurate tongue placement with centralized airflow. In addition to having worse speech and language outcomes than other phenotypic subgroups, participants with microtia plus mandibular hypoplasia also had the highest rate of failed hearing screens. Therefore, speech and language deficits may be more attributable to hearing impairment, as has been documented in non-CFM populations (Anne et al.,

Table 6. Comparison of mean speech and language scores for adolescents with and without craniofacial microsomia.

Test and measure	IPW					Excluding Spanish-speaking participants ^a					CNR analyses			
	Mean diff. ^c	95% CI	ES ^d	<i>p</i> value		Mean diff. ^c	95% CI	ES ^d	<i>p</i> -value		Mean diff. ^c	95% CI	<i>p</i> value	
CELF ^a														
Recalling Sentences	-1.23	-2.01	-0.44	-0.43	.002	-0.66	-1.36	0.04	-0.24	.06	-2.45	-3.40	-1.50	< .001
Receptive	-1.26	-2.05	-0.46	-0.46	.002	-0.35	-1.02	0.32	-0.13	.30	-2.17	-3.13	-1.21	< .001
Expressive	-0.90	-1.70	-0.09	-0.30	.03	-0.33	-1.03	0.36	-0.12	.34	-2.16	-3.13	-1.20	< .001
CCC2-Parent ^{a,b}														
Speech	-1.14	-1.97	-0.32	-0.40	.01	—	—	—	—	—	-3.24	-4.30	-2.19	< .001
Syntax	-0.13	-0.79	0.53	-0.06	.69	—	—	—	—	—	-1.88	-2.73	-1.04	< .001
Semantics	0.08	-0.57	0.73	0.03	.81	—	—	—	—	—	-1.73	-2.56	-0.90	< .001
Coherence	-0.15	-0.87	0.56	-0.06	.67	—	—	—	—	—	-2.20	-3.11	-1.28	< .001
Nonverbal Comm.	-0.42	-1.06	0.21	-0.19	.19	—	—	—	—	—	-2.05	-2.92	-1.19	< .001
General Comm.	-1.50	-5.50	2.51	-0.10	.46	—	—	—	—	—	-12.37	-17.44	-7.30	< .001
CCC2-Teacher ^a														
Speech	-1.18	-2.08	-0.27	-0.42	.01	-1.65	-2.49	-0.82	-0.60	< .001	-3.47	-4.75	-2.19	< .001
Syntax	-0.26	-1.03	0.52	-0.10	.52	-0.72	-1.37	-0.07	-0.30	.03	-2.64	-3.68	-1.60	< .001
Semantics	-0.59	-1.34	0.15	-0.24	.12	-0.92	-1.59	-0.26	-0.38	.01	-2.70	-3.70	-1.71	< .001
Coherence	-0.01	-0.80	0.78	0.00	.98	-0.49	-1.22	0.23	-0.19	.18	-2.36	-3.43	-1.29	< .001
Nonverbal Comm.	-0.63	-1.40	0.14	-0.26	.11	-1.28	-2.03	-0.53	-0.52	.001	-3.07	-4.19	-1.95	< .001
General Comm.	-2.48	-6.84	1.88	-0.17	.27	-5.49	-9.42	-1.55	-0.40	.01	-15.05	-20.92	-9.19	< .001
TDTA														
Percent correct	-5.63	-7.81	-3.46	-1.12	< .001	-5.14	-7.31	-2.97	-1.02	< .001	-9.18	-12.53	-5.82	< .001
SIT														
Percent correct	-5.44	-8.54	-2.33	-0.77	.001	-3.57	-5.96	-1.18	-0.59	.004	-8.73	-13.40	-4.06	< .001

Note. Em dashes indicate data not applicable. IPW = inverse probability weighting; CNR = censored normal regression; diff. = difference; CI = confidence interval; CELF = Clinical Evaluation of Language Fundamentals; CCC-2 = Children's Communication Checklist-2; Comm. = Communication; TDTA = Templin–Darley Tests of Articulation; SIT = Speech Intelligibility Test.

^aScaled scores used for all CELF and CCC-2 measures, with the exception of the CCC-2 General Communication Composite, which uses standard scores ($M = 100$, $SD = 15$). ^bSpanish-speaking parents did not complete a CCC-2. ^cAdjusted for age (continuous), gender, race (White/non-White), income (categorical), and primary caregiver's highest level of education (categorical). ^dES = standardized effect size, not calculated for CNR analyses.

2017), which disproportionately affects children with mandibular hypoplasia plus microtia. Ideally, future studies would include serial audiological evaluations in addition to phenotypic assessment in order to determine potential targets for intervention in this population.

Youth with CFM tended to score lower than their peers on ratings of communication behaviors, particularly those based on teachers' rather than parents' reports. These included elements of communication that are heavily influenced by speech intelligibility (e.g., articulation), as well as nonverbal communication. These differences in verbal and nonverbal communication may be exacerbated by the stigma associated with facial, speech, or vocal quality differences. Similar "social inhibition" has been reported in other craniofacial populations (Padwa, Evans, & Pillemer, 1991; Pertschuk & Whitaker, 1985). We observed a similar finding in this cohort in the early elementary school years, when teachers' reports suggested that children with CFM exhibited greater problems with anxiety and social function than unaffected peers (Dufton et al., 2011). Early speech-language therapy may help to attenuate these differences, by instilling in young children increased confidence in their speech and by addressing the pragmatic and nonverbal elements of communication.

Clinically, our findings argue for management of hearing loss (e.g., use of hearing aids, classroom accommodations

as needed) and monitoring of speech and language function in children with CFM as part of their craniofacial care. Our impression is that clinical practice standards in both respects are highly variable, in contrast, for example, to guidelines of care for children with cleft palate. In addition to facilitating timely referral for speech-language therapy and hearing services, more consistent and focused speech and language monitoring may help to inform other aspects of the child's care (e.g., timing of surgeries to improve mandibular function, possible re-animation procedures for children with cranial nerve deficiency).

Like most initial studies in populations with rare conditions, this research had several limitations. CFM is an inherently heterogeneous condition, and research examining CFM features in relation to functional outcomes is lacking. Although our phenotypic classifications were based on a robust and reliable method for categorizing CFM phenotype, further research is needed to examine more granular characteristics that might be most closely related to developmental outcomes. Ideally, this would include replication of our results in studies designed to examine hypothesized associations between CFM characteristics and functional outcomes (e.g., mandibular hypoplasia, jaw range of motion, and speech sound production). Second, we used an abbreviated assessment battery to characterize

speech and language skills, which may have missed more nuanced group differences that could affect communication. We have minimal data on the number of youth who exhibited early speech delays that may have resolved later or youth who developed compensatory strategies. While our findings provide justification for further assessment and monitoring of speech and language in children and youth with CFM, replication with a more detailed assessment battery in younger children is needed to identify specific targets for intervention. Although we had some data on the number of participants who had received speech-language intervention, we did not collect detailed information on the type, intensity, or duration of services received. Thus, for example, while a high proportion of youth with CFM (i.e., 60%) received at least some speech-language intervention, we do not know how many received “adequate intervention” in terms of frequency and duration. Finally, our hearing screenings were conducted in a variety of test settings (homes, libraries, etc.), which were not always optimal in terms of background noise. Further, a single-hearing screening cannot capture fluctuations in hearing or hearing impairment at previous critical developmental periods. Although this was an improvement over reliance on parent reports or review of medical records, this method is not equivalent to a detailed audiological assessment, which would provide information on both type and degree of hearing loss. For example, we may have missed instances of mild or transient hearing loss or the rate of hearing impairment may have been inflated due to testing in suboptimal conditions. This would have affected both groups. If anything, such misclassification would attenuate our estimates of the associations between hearing loss and speech, language, and communication measures. Ideally, youth with CFM would undergo complete audiological assessments and appropriate follow-up to minimize the impact of hearing impairment on communication skills.

These findings argue for routine assessment of speech and language function in children with CFM, particularly those with mandibular hypoplasia and microtia and those with hearing loss. Although international standards of care specific to CFM do not exist, craniofacial centers providing team care have the expertise to assess speech and language concerns and hearing loss in this population. Further research, ideally using prospective assessments of speech, language, and hearing beginning in early childhood, is needed to identify specific targets for assessment and intervention.

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