Psychosocial Outcomes in Children with Hemifacial Microsomia

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Objective To determine whether children with hemifacial microsomia (HFM) have higher risk for psychosocial problems than children without HFM. Methods One hundred and thirty-six children with HFM (64% male, mean age = 6.9 years) were compared to 568 matched controls (50% male, mean age = 7.0 years) on parent and teacher measures of behavior problems and social competence, and teacher rankings of peer acceptance. Results Parents of cases and controls reported similar levels of behavior problems and social competence. Teachers reported higher frequencies of internalizing problems, lower social competence and less peer acceptance for cases. Relative to controls, teacher-rated outcomes were worse for female cases, those with younger mothers at the time of birth, those with eye anomalies, and those with one or more malformations in addition to the core features of HFM. Conclusions This study provides the first evidence of relatively poor psychosocial outcomes among children with HFM.

Key words children; craniofacial; Goldenhar syndrome; ocularauricular vertebral syndrome; psychosocial.

Introduction

Hemifacial microsomia (HFM) is a variable, complex congenital disorder that affects the development of craniofacial structures including the condyle and ramus of the mandible, zygomatic arch, malar bone, external ear, middle ear ossicles, temporal bone, cranial nerves, and muscles of facial expression (Gorlin, Cohen, & Hennekam, 2001; Lauritzen, Lilja, & Jarlstedt, 1986; Poswillo, 1973). In addition to these anomalies affecting the ear, eye, jaw, and cheek, HFM is associated with a variety of other malformations such as oral clefts and defects of the heart, kidney, and limbs (Gorlin et al., 2001; Poswillo, 1973). Children with HFM have an elevated risk of functional impairments in vision, hearing, speech, feeding/eating, and, if severe, breathing due to tracheal obstruction (Stromland et al., 2007). The term “hemifacial” refers to the typical asymmetry of affected tissues, but under- or mal-development can be bilateral. The term “craniofacial microsomia” is sometimes used to acknowledge this bilaterality.

HFM is the second most common craniofacial birth defect, with a prevalence of 1 in 3,500–18,000 births (Araneta et al., 1997; Grabb, 1965). Pediatric psychologists in hospital settings are therefore likely to encounter these children, particularly in light of the American Cleft Palate-Craniofacial Association’s practice parameters that require craniofacial teams to provide psychological assessment and intervention services (American Cleft Palate-Craniofacial Association, 1993, 1996). However, little is known about the psychological correlates and outcomes of HFM. Relevant studies have been limited by diagnostically heterogeneous samples containing only a small number of children with HFM. These investigations have shown that children with various craniofacial disorders are
often more inhibited, depressed, anxious, introverted, and less socially adept than typical children, as indicated by standardized behavior checklists and interviews (Padwa, Evans, & Pillemer, 1991; Pertschuk & Whitaker, 1985; Pillemer & Cook, 1989). In one study, investigators found a trend for a subgroup of children with HFM to have higher anxiety and lower self-concept scores than children with other craniofacial conditions (Pertschuk & Whitaker, 1985). Padwa et al. (1991) found that one third of the children with HFM in their sample had scores suggesting clinically significant levels of behavior problems in relation to normative data, but Snyder and Pope (2010) showed that the 47 children with HFM in their sample were well adjusted in relation to behavior checklist norms. Fewer than 100 HFM cases have been examined across all studies, limiting the extent to which findings can be generalized to the broader population of HFM-affected children. Further, most prior investigations used test norms rather than demographically similar control groups. The relative psychosocial status of children with HFM therefore remains uncertain.

The current study was designed to test the hypothesis that young children with HFM are more likely to experience psychosocial difficulties than demographically similar children without HFM. This expectation follows from the previously cited studies of children with craniofacial disorders including HFM, as well as studies of other children sharing one or more of the risk factors commonly found in HFM. These include anomalous or unattractive craniofacial appearance, which can lead to poor social acceptance by peers (Dion & Berscheid, 1974; Gertner, Rice, & Hadley, 1994). In children with HFM, this effect that may be exacerbated by cranial nerve defects and associated limitations in facial expressiveness or eye gaze (Bogart & Matsumoto, 2010). The hearing, vision, and speech impairments that often accompany HFM have been generally linked to psychiatric disorders, poor behavioral functioning, and poor social communication (Brown, Hobson, Lee & Stevenson, 1997; Fellinger, Holzing, Sattel, Lauchet, & Goldnerg, 2009; McAlpine & Moore; 1993; van Daal, Verhoeven & van Balkom, 2007). We have previously shown that HFM-affected children have elevated risk for receptive language and visual-motor problems (Collett et al., 2010), further supporting our hypothesis that psychosocial difficulties may be more common in HFM because neurocognitive and psychosocial deficits have been shown to cluster in other samples of children (Coleman & Minnett, 1993).

We therefore postulated that children with HFM would have higher levels of behavior problems, less social competence and less acceptance by their classroom peers than control group children. These hypotheses were addressed by evaluating psychosocial outcomes in a cohort of 136 school-age children with HFM whose mothers had participated in a study of pregnancy risk factors when the child was younger than 3 years (Werler et al., 2004). Cases were compared to a control group of 568 children without known structural malformations, who were recruited at the same time as cases and case-matched by age and pediatrician. In secondary analyses, we examined differences among subgroups of children with HFM and as a function of demographic characteristics. Case subgroups were identified in relation to distinct combinations of the “core malformations” that define HFM (i.e., ear, eye, jaw, and cheek anomalies). We examined these combinations to see if any have worse outcomes, as this issue has not been studied previously and there is little information upon which to base hypothesized associations with psychosocial outcomes. We also identified case subgroups in relation to the presence of one or more “other malformations” such as cleft lip/palate and cardiac or renal defects. Here we anticipated worse outcomes among cases with one or more other malformations, due to cumulative risk (i.e., these malformations introduce additional surgeries and functional limitations). We also examined whether child gender or maternal age would modify the results, based on well-known gender differences in the prevalence of behavior problems (i.e., boys tend to have more externalizing behavior, girls are more prone to internalizing behavior; Costello, Compton, Keeler, & Angold, 2003), and previous studies suggesting that young maternal age is associated with worse psychosocial outcomes for children (Fergusson, Horwood, & Lynskey, 1993; Furstenberg, Brooks-Gunn, & Chase-Lansdale, 1989).

In summary, the present study expanded upon previous psychosocial studies of HFM in several ways, including: the recruitment of a relatively large group of cases, all having HFM; comparisons with a demographically matched control group; reports of outcomes by parents and teachers; and the examination of outcomes by subgroups distinguished by phenotype or selected demographics. We previously published a preliminary report on teacher reports of psychosocial functioning for a subset of the study sample (Werler, Starr, Cloonan, & Speltz, 2009). In addition to including the full sample of cases and controls, the current study added parents’ reports of functioning, teachers’ rankings of peer acceptance and analyses that more rigorously controlled for potential confounders and examined subgroup differences among cases.

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Methods
Participants
Participants from the Werler et al. (2004) study were re-approached to participate in the current study when their child was 5–12 years old.

Children with HFM
The mothers of 279 children with HFM were originally enrolled from 26 craniofacial centers across the United States and Canada (Werler et al., 2004). Mothers were eligible to participate if their affected child was <4 years old, had not been adopted, and had no known chromosomal defects. The present follow-up study of these children did not include 12 cases due to institutional review board constraints. For this analysis, 62 children were not included for diagnostic reasons (40 with unilateral microtia and no other evidence of HFM, 11 twin gestations, 5 with isotretinoin-related HFM, and 6 with genetically inherited HFM). Another nine participants were excluded due to family circumstances (e.g., child death, out of home placement). Of the remaining 196 eligible mother/child pairs, 43 could not be contacted and 17 refused participation, resulting in a final sample of 136 cases (69% of eligible case participants).

Unaffected Controls
The mothers of 884 unaffected controls were enrolled in the original study (Werler et al., 2004). Eligible controls had no known birth defects, had not been adopted, and were recruited for each case through cases’ pediatricians or from another pediatric practice of approximately the same size as and in close proximity to cases’ pediatricians. In the original cohort study, approximately three controls were matched to each case for age (±2 months of case’s age at the time of recruitment) and pediatric practice. Due to institutional review board constraints, 45 controls were not included in the follow-up study. For this analysis, six control families were excluded due to family circumstances and two were excluded due to twin gestation. Of the remaining 831 families, 142 could not be contacted and 121 refused participation, resulting in a final sample of 568 controls (68% of eligible controls).

Measures
Medical Records Review
Medical records for cases were reviewed for notations of anomalous structures, including the core components of HFM (i.e., malformations of the ear, eye, jaw, and cheek) and other major malformations. Cases with “other” malformations (HFM + Other) included cleft lip and/or palate; defects of the heart, kidney, limb, intestines, and genitals; and Larson’s or Moebius Syndrome (Table I). These cases were distinguishable from “isolated” cases having one or more of the core features of HFM, but no other documented conditions or syndromes. Medical records were unavailable for eight cases.

Demographic and Medical History Interview
Participating mothers were interviewed by a research nurse on demographic, reproductive, medical, and behavioral factors (see Werler et al., 2004). Data utilized for this study included race/ethnicity, maternal age at the time of the child’s birth, maternal education, household income, marital status, language spoken in the home, and geographic region of residence.

Child Behavior Checklist
The Child Behavior Checklist (CBCL; Achenbach & Rescorola, 2001) provides a measure of parent-reported externalizing behavior problems (e.g., hyperactive, noncompliant, disruptive) and internalizing behavior problems (e.g., shy, withdrawn, despondent). The current study used three broadband scales from the CBCL (Internalizing, Externalizing, and Total Problems) and eight syndrome scales (Aggressive Behavior; Anxious/Depressed; Attention Problems; Rule-Breaking Behavior; Social Problems; Somatic Complaints; Thought Problems; and Withdrawn/Depressed). T-scores are derived for each domain (Mean = 50, SD = 10), based on separate norms for boys and girls from children drawn from a nationally representative sample. Spanish-speaking parents were administered the Spanish version of the CBCL (Achenbach et al., 1998). Internal consistency (Cronbach’s α) for the CBCL broadband scales ranged from α = .79 to .94. Internal consistency for syndrome scales ranged from α = .60 to .86.

Teacher Report Form
The Teacher Report Form (TRF) is the companion instrument to the CBCL and is designed to obtain teachers’ reports of children’s behavioral and emotional problems (Achenbach & Rescorola, 2001). The TRF provides T-scores for the same three broadband and eight syndrome scales listed above for the CBCL. Internal consistency for the TRF broadband scales ranged from α = .79 to .96, and for the syndrome scales ranged from α = .52 to .92.

Social Competence Scale—Parent Version
The Social Competence Scale—Parent Version (SCP) is a 12-item measure that assesses a child’s prosocial behaviors, communication skills, and self-control (Conduct Problems Prevention Research, 1999). Each item on this scale describes a behavior that a child may display in a social
setting (e.g., “Your child can give suggestions and opinions without being bossy” and “Your child can calm down when excited or all wound up.”). Parents rate how well each statement describes their child using a 5-point Likert scale ranging from 0 (Not at all) to 4 (Very Well). The SCP contains two subscales: Prosocial/Communication Skills and Emotional Regulation Skills. A total score is also reported. Subscale and total scores are based on the means of the relevant item ratings. Investigators have found that SCP subscale and total scores distinguished high-risk early grade school children from control groups, with good internal consistency of the scales (Conduct Problems Prevention Research Group, 1999). A Spanish translation of the SCP was provided by Language Line Document Translation, a division of Language Line Services of California. Internal consistency for the SCP for this sample was $\alpha = .90$.

### Social Competence Scale—Teacher Version

The Social Competence Scale—Teacher Version (SCT) is the companion instrument to the SCP. The SCT is a 25-item measure that assesses a child’s prosocial and communication skills, emotional self-regulation, and academic skills using the same item format and scoring procedures and similar subscales. One difference is an additional subscale, Academic Skills. Like the SCP, SCT subscale scores, the combined score, and the total score are calculated as the mean of responses. This instrument has also shown good reliability and validity (Conduct Problems Prevention Research Group, 1999). Internal consistency of the SCT for this sample was $\alpha = .97$.

### Teacher Peer Acceptance Rankings

Teacher rankings of sociometric status were used to estimate the social acceptance of cases and controls by their peers (Peer Acceptance Ranking; PAR). Teachers were asked to rank order all children in their classroom from the most to least accepted. The numeric position of the child participating in this study was identified and expressed as a percentage of the total number of children in a given classroom of the same gender; higher percentage scores indicated less peer acceptance. In previous research involving similarly aged children, teacher rankings of peer acceptance have correlated positively with children’s own and same-aged peer rankings of acceptance (Matson, Esveldt-Dawson, & Kazdin, 1983; Wu, Hart, Draper, & Olsen, 2001).

### Procedure

Mothers were initially contacted by phone, and those who expressed interest in participating were mailed a packet...
containing the consent form and study questionnaires. For Spanish-speaking families, translations of the parent measures (CBCL, SCP) were provided. Mothers who consented to participate identified a teacher who could complete questionnaires. For children who were home-schooled \((n = 19)\), teacher questionnaires were not completed. For some children \((n = 16)\), a classroom teacher was unavailable, and families identified a similarly trained adult (e.g., a speech therapist, school counselor) familiar with the child who completed the “teacher” questionnaires. For these children, a PAR form was not completed because these adult raters did not typically observe the child in a classroom setting. The child’s teacher or other school staff member was sent a packet including the TRF, SCT, and PAR.

This study was approved by the sponsoring institution’s Institutional Review Board and was completed in full compliance with HIPAA standards.

**Data Analysis**

Demographic and clinical characteristics of HFM cases and controls were summarized using descriptive statistics. The primary hypothesis, which was that children with HFM were more likely to experience psychosocial difficulties than controls, was evaluated using multiple linear regression, adjusted for the following demographic characteristics: gender, race/ethnicity, maternal age, maternal education, family income, Spanish language, study center, and marital status. To determine the magnitude of group differences after adjusting for these covariates, standardized mean difference effect sizes (ES) were calculated using a modification of Cohen’s \(d\) (Cohen, 1988) by taking the adjusted difference in means and dividing by the root mean square error for the model. Using Cohen’s (1988) criteria, we defined effect sizes less than 0.2 as “negligible,” those between 0.2 and 0.5 “small,” those between 0.5 and 0.8 “medium,” and those greater than 0.8 as “large.”

In order to estimate the relative status of cases and controls in relation to clinical cut-off scores, odds ratios (ORs) with 95% confidence intervals (CIs) were calculated to compare the proportion of cases and controls in the “borderline clinical” and “clinical” range on the CBCL and TRF, defined by Achenbach and Rescorla (2001) as a \(T\)-score of 60 or greater for the broadband scores and 65 or greater for the syndrome scales.

To evaluate hypotheses regarding potential effect modification, we used multiple regression to examine case-control group differences separately by child gender and categories of maternal age at the time of the child’s birth \((\leq25, >25)\), controlling for the confounders listed above (except that a confounder was excluded when examining subgroups based on that variable). We similarly compared controls on all outcome measures to: (a) cases with additional malformations (HFM + Other) and (b) seven case subgroups, each representing one of the possible combinations of ear, eye, and jaw/cheek anomalies (e.g., ear only, ear + eye, ear + eye + jaw/cheek). As detailed in the results, we collapsed across combinations having too few cases to analyze separately.

In sensitivity analyses, we used inverse probability weighting (IPW) for all analyses to estimate the effects of response bias on all dependent variables. IPW is commonly used to adjust for confounding and selection bias associated with variables measured at earlier time points (Heyting, Tolboom, & Essers, 1992). For these analyses, logistic regression was used to predict participation in a given outcome measure using the demographic variables and other covariates listed above. Scores were then generated to reflect the probability of each participant being observed (i.e., participating) on the given measure. Linear regression analyses were re-run, weighted by the inverse of the probability of being observed, to determine whether there was a meaningful change in results. For example, if children of unmarried mothers were only half as likely to participate as the children of married mothers, the scores generated by the children of unmarried mothers who actually participated would be weighted by a factor of 2. Note that this analysis accounts for both non-participation in follow-ups (either because of inability to contact or refusal) as well as the “missingness” of specific outcomes among those who participated in some assessments.

As this is the first large-scale study of psychosocial functioning in children with HFM in relation to a control group, we did not want to miss potentially important group differences that would warrant future study (i.e., Type II errors). We therefore did not adjust \(p\)-values for multiple comparisons in these exploratory analyses; nor did we interpret \(p\)-values as dichotomous significance tests. Rather, we evaluated group differences in terms of the magnitude of the associations observed and the precision of such estimates (see Perneger, 1998 for further justification for this approach).

**Results**

Participants in the follow-up assessments were similar to non-participants from the original cohort in child gender. However, there were substantial differences in maternal demographic characteristics, including Hispanic ethnicity (14 vs. 31% for participants and non-participants,
respectively), less than high school education (11 vs. 30%), family income $25,000 or less (18 vs. 36%), unmarried mother (22 vs. 38%), and average maternal age (29 vs. 27 years). Among participants, rates of complete data records were high, ranging from approximately 88% for TRF and PAR and 91% for SCT to 99-100% for SCP and CBCL. The application of IPW to adjust for bias associated with participation and incomplete data records did not appreciably alter any of the findings. For all measures, the magnitude of group differences was either nearly identical or somewhat larger with the application of IPW (data not shown). As a result, only unweighted results are presented below.

Table I summarizes the demographic characteristics of the entire sample and, for cases, significant clinical features. Group differences between cases and controls on age (cases mean = 7.0 years, SD = 1.0; controls mean = 6.9 years, SD = 1.0), socioeconomic status, or geographic location were minimal. There were more males in the case group (64% vs. 50% in controls). More cases were of Hispanic heritage (24% vs. 12% in controls) and came from Spanish-speaking households (12% vs. 3% in controls). Home schooling was infrequent in both groups (<1% cases, 2% controls) and most children were in kindergarten through second grade (81% cases, 83% controls).

Combinations of core anomalies that constitute HFM were distributed as follows: 4 with ear anomalies only, 1 with eye only, 12 with jaw/cheek only, 1 with ear + eye, 78 with ear + jaw/cheek, 4 with eye + jaw/cheek, 20 with ear - eye + jaw/cheek, and 8 whose medical records lacked sufficient detail for classification. Forty-six (34%) cases were in the HFM + Other group, and had one or more of the following conditions: cleft lip and/or palate (n = 14); cardiac (n = 26), renal (n = 6), limb (n = 7), gastrointestinal (n = 8) and genital malformations (n = 6); Moebius (n = 1) and Larsen’s syndrome (n = 1).

### Comparison of Cases and Case Subgroups with Controls

**Behavior Problems Reported by Parents**

No group differences on the internalizing, externalizing, and total scales for the CBCL were found (Table II). On the CBCL syndrome scales (Table III), differences were negligible with most effect sizes ranging from −0.11 to 0.17 (p = 0.087–0.995). On the social problems scale cases scored higher than controls (p = 0.002), but the effect size was small (ES = 0.30). These findings were not altered by the different combinations of core malformations, the presence of other malformations, child gender, or mothers’ age at birth.

**Behavior Problems Reported by Teachers**

On the TRF, teachers rated children with HFM as having higher levels of internalizing, externalizing, and total behavior problems than controls, with negligible to small differences (ES = 0.16–0.27, p = 0.005–120) (Table II). Teachers rated cases as having more difficulties than controls on the

### Table II. Case–Control Comparisons on Broadband Indices of Psychosocial Functioning

<table>
<thead>
<tr>
<th>Measure</th>
<th>No. Cases/Controls</th>
<th>Cases Mean</th>
<th>SD</th>
<th>Controls Mean</th>
<th>SD</th>
<th>Differencea</th>
<th>95% CI</th>
<th>p</th>
<th>ES</th>
</tr>
</thead>
<tbody>
<tr>
<td>TRF: Internal</td>
<td>135/564</td>
<td>49.70</td>
<td>9.17</td>
<td>48.26</td>
<td>9.68</td>
<td>0.34</td>
<td>−1.50, 2.17</td>
<td>.718</td>
<td>0.04</td>
</tr>
<tr>
<td>TRF: External</td>
<td>135/564</td>
<td>48.50</td>
<td>9.26</td>
<td>47.77</td>
<td>9.77</td>
<td>0.42</td>
<td>−1.45, 2.29</td>
<td>.657</td>
<td>0.04</td>
</tr>
<tr>
<td>TRF: Total</td>
<td>135/564</td>
<td>49.44</td>
<td>9.20</td>
<td>47.22</td>
<td>10.45</td>
<td>−0.19</td>
<td>−0.34, −0.03</td>
<td>.36</td>
<td>−0.02</td>
</tr>
<tr>
<td>SCP: Emotional</td>
<td>136/568</td>
<td>2.27</td>
<td>0.76</td>
<td>2.30</td>
<td>0.71</td>
<td>−0.01</td>
<td>−0.13, 0.13</td>
<td>.881</td>
<td>−0.01</td>
</tr>
<tr>
<td>SCP: Communication</td>
<td>135/568</td>
<td>2.80</td>
<td>0.74</td>
<td>2.83</td>
<td>0.72</td>
<td>−0.002</td>
<td>−0.14, 0.01</td>
<td>.981</td>
<td>0.00</td>
</tr>
<tr>
<td>SCP: Total</td>
<td>136/568</td>
<td>2.53</td>
<td>0.68</td>
<td>2.56</td>
<td>0.66</td>
<td>−0.006</td>
<td>−0.13, 0.12</td>
<td>.921</td>
<td>−0.01</td>
</tr>
<tr>
<td>SCT: Emotional</td>
<td>126/511</td>
<td>2.72</td>
<td>0.87</td>
<td>2.83</td>
<td>0.80</td>
<td>−0.15</td>
<td>−0.31, 0.01</td>
<td>.069</td>
<td>−0.18</td>
</tr>
<tr>
<td>SCT: Communication</td>
<td>126/511</td>
<td>2.83</td>
<td>0.85</td>
<td>3.00</td>
<td>0.78</td>
<td>−0.22</td>
<td>−0.38, −0.07</td>
<td>.005</td>
<td>−0.19</td>
</tr>
<tr>
<td>SCT: Academic</td>
<td>126/514</td>
<td>2.53</td>
<td>1.07</td>
<td>2.71</td>
<td>0.92</td>
<td>−0.20</td>
<td>−0.39, −0.01</td>
<td>.036</td>
<td>−0.21</td>
</tr>
<tr>
<td>SCT: Total</td>
<td>126/513</td>
<td>2.70</td>
<td>0.88</td>
<td>2.85</td>
<td>0.78</td>
<td>−0.19</td>
<td>−0.34, −0.03</td>
<td>.019</td>
<td>−0.24</td>
</tr>
<tr>
<td>PAR</td>
<td>121/496</td>
<td>51.59</td>
<td>22.85</td>
<td>43.33</td>
<td>21.64</td>
<td>9.39</td>
<td>4.84, 13.94</td>
<td>.000</td>
<td>0.43</td>
</tr>
</tbody>
</table>

Note. Measures include the CBCL, TRF, SCP, SCT, and PAR.

TRF and CBCL: T-score.

aDifferences between groups after adjusting for: child’s gender, child’s age, family income, parent’s marital status, mother’s age at birth, mother’s years of education, mother’s race/ethnicity, English spoken at home (yes/no), and region of country.
following TRF syndrome scales (Table III): withdrawn/depressed (ES = 0.27), somatic complaints (ES = 0.21), social problems (ES = 0.29), attention problems (ES = 0.24), and aggressive behavior (ES = 0.21).

With respect to effect modifiers of TRF scores, internalizing, externalizing and total behavior problems were higher for female cases than for female controls (ES = 0.29), attention problems (ES = 0.24), and aggressive behavior (ES = 0.21). Compared to controls, teachers rated HFM + Other cases as having higher levels of internalizing (ES = 0.32) and total problems (ES = 0.37). When case children were grouped by core malformations (both single anomalies and combinations of two or more), mean scores were higher than controls for 3 of these subgroups: (a) eye + jaw/cheek, (b) ear + eye, and (c) eye + ear + jaw/cheek. Because the number of children in each of these groups was small, and they each involved eye anomalies plus at least one other core anomaly, we combined them (n = 25), allowing us to control for demographic factors when estimating differences in scores. In these analyses, teachers were found to rate this eye anomaly subgroup as having higher internalizing behavior scores (ES = 0.40) and higher total behavior scores (ES = 0.40) compared to controls. Alpha levels for all analyses of effect modifiers were less than 0.05, with the exception of the internalizing behavior finding for cases with eye anomalies (p = .14).

When examining scores categorically, OR and 95% CIs showed children with HFM were twice as likely as controls to receive scores in the “borderline clinical” range or higher on the TRF Internalizing (OR = 2.27, 95% CI 1.14–4.52), Externalizing (OR = 2.93, 95% CI 1.52–5.67), and Total broadband scales (OR = 2.57, 95% CI 1.38–4.78). Children with HFM were six times as likely as controls to receive scores in the “clinical” range on the TRF social problems syndrome scale (OR = 6.18, 95% CI 2.44–15.69). They showed elevated risk on TRF syndrome scales for withdrawn/depressed (OR = 3.89, 95% CI 1.42–10.65); thought problems (OR = 2.56, 95% CI 1.03–6.33); attention problems (OR = 2.57, 95% CI 1.04–6.32); and rule-breaking behavior (OR = 3.93, 95% CI 1.61–9.62). In addition, there was evidence that cases were more likely to be anxious/depressed (OR = 1.47, 95% CI 0.58–3.69); have somatic complaints (OR = 2.33, 95% CI 0.70–7.77); and be aggressive (OR = 2.75, 95% CI 0.88–8.59), but these ORs were smaller and less precise.

### Social Competence Reported by Parents

Case-control differences on the SCP were small in magnitude and imprecise (ES = 0.00 to −0.01, p = .881–.981) (Table II).
Social Competence Reported by Teachers
Teachers rated cases as having lower overall social competence (ES = −0.24) and poorer prosocial/communication (ES = −0.19) and academic skills (ES = −0.21) than controls (Table II). Case–control differences on the social competence total were largest for female cases (ES = −0.35) and cases with younger mothers (ES = −0.70). The magnitude of group differences in teacher-reported social competence was unaffected by the various combinations of core malformations and the presence of other malformations.

Teacher Rankings of Peer Acceptance
The mean percentage ranking for cases as rated by teachers was 52% compared to 42% for controls (Table II), indicating lower peer acceptance for cases on average. This effect held for males and females, both maternal age groups, all combinations of core malformations and HFM + Other cases. We further examined individual peer acceptance rankings by specific type of other malformation. Among the 14 cases with oral clefts in our sample, 9 received peer acceptance rankings below the mean acceptance ranking for all cases; eight of these 9 children also had ear and jaw/cheek anomalies.

Discussion
Despite the many functional correlates of HFM that complicate psychosocial adjustment such as hearing and speech problems and anomalous facial appearance, there has been little study of this disorder from a psychological perspective. The present study is the first to examine the psychosocial characteristics of a large sample of children with HFM in relation to a comparison group allowing for control of sociodemographic variables that potentially influence psychosocial outcomes. Given that cases and controls were drawn from 26 different communities across the United States and Canada, we relied on parents’ and teachers’ questionnaire responses to assess children’s psychological functioning.

Differences Between Cases and Controls
Our central hypothesis—that cases would fare worse than controls—was partially supported. Although teachers rated children with HFM as having more behavior problems and social difficulties, parents of cases and controls provided nearly equivalent ratings of their children’s behaviors in these areas. The only exception for parents was a higher average score for cases on the social problems scale of the CBCL. However, group differences on this scale may have been inflated by the inclusion of one item (“speech problems”), an area of known difficulty for children with HFM (Collett et al., 2010). This serves as a reminder that the CBCL and other standardized instruments can sometimes lead to spuriously high estimates of behavioral disturbance in children with medical conditions (Perrin, Stein, & Drotar, 1991).

The contrast in this study between parent and teacher reports is not surprising, given the extensive literature suggesting that cross-informant agreement on these and similar scales is low to moderate (Achenbach, 1991; Frigerio et al., 2004; Mitsis, McKay, Schulz, Newcorn, & Halperin, 2000; Salbach-Andrae, Lenz, & Lehmkuhl, 2009). The pattern of disagreement among different informants has varied considerably in these studies, depending on child age and culture, and some studies have found teachers reporting fewer behavior problems than parents (Youngstrom, Loeb, & Southamer-Lober, 2000). In the current study, teachers may have been more aware than parents of problems among cases due to the social context in which they observed children. Peer environments may be particularly difficult for children with HFM due to speech or hearing deficits, inhibited behavior, or anomalous facial appearance. These factors and any associated inhibited behaviors may exert less influence in the context of close, family relationships. Certain reporting biases may have also contributed to the discrepancy in caregiver reports. Parents may tend to minimize problems in their optimistic view of the child’s status, and teachers may be susceptible to the well-studied social bias associated with unattractive or anomalous faces (Langlois et al., 2000; Tobiasen, 1987). In future studies of this population, as well as clinical work, child-report measures, including interviews, should be used to obtain the child’s perspective, which may provide a better account of problem behaviors and competencies than those taken from adults, especially internalizing symptoms and other subjective experiences (Long, Forehand, & Wiers, 1992).

The magnitude of teacher-reported group differences was relatively modest on nearly all measures and scales. Although teacher rankings of peer acceptance indicated that cases lagged nearly 10 percentage points behind their peers, both groups were perceived by their teachers as having social status close to the middle of their respective classroom groups. This is remarkable, given the anticipated negative effects of anomalous appearance and poor speech on peer popularity (Dion & Berscheid, 1974; Gertner et al., 1994). Ratings by the students themselves are needed to confirm this impression based on adult perceptions.

Similarly, cases were more than twice as likely as controls to have TRF standard scores in the clinical range, but
the absolute number of children falling into this range was relatively low for both groups (e.g., on the TRF social problems scale, approximately 13% of cases vs. 6% of controls had T scores in the borderline clinical range). Children with extreme levels of social rejection or problem behavior were therefore rare among those included in the study. This may point to the resilience of children with HFM (Snyder & Pope, 2010), but more likely reflects the demographically stable, low-risk nature of the sample itself (e.g., 93% of our families had two parents; nearly half contained mothers with college degrees). We would anticipate higher levels of clinically significant problems among both cases and controls in samples more closely matched to the population in their representation of socioeconomic risk (Costello, Compton et al., 2003; Cuffe, McKeown, Addy, & Garrison, 2005).

Comparison of Case Subgroups with Controls

Although parent measures did not differ by case subgroup, teacher reports followed our expectations on some measures. Teachers rated cases with other malformations (particularly oral clefts) and cases with eye anomalies—in combination with either or both ear and jaw/cheek anomalies—as having worse outcomes than controls (i.e., more behavior problems and less peer acceptance). Unfortunately, our medical records lacked the specificity and standardization to narrow down the range of causal pathways that might account for these associations. For example, observed eye anomalies included a mix of dermoids, colobomas, eyelid anomalies (nerve or tissue), and small orbits. The effect of such anomalies on psychosocial outcomes could reflect vision impairments, social bias in response to craniofacial appearance, or eye movements that facilitate reciprocal social interactions (e.g., eye contact, symmetrical eye movements). Similarly, our measure of peer acceptance did not allow for identification of specific social difficulties encountered by these children such as peer rejection or neglect (Hoglund, Lalonde, & Leadbeater, 2008). Future research is needed to test the hypothesis that eye and certain other malformations carry greater risk, using methods that: (a) more precisely distinguish between specific anomalies and associated functional impairments and (b) more specifically measure peer group processes. In the meantime, our findings suggest that children with HFM who have eye anomalies in combination with other core features of HFM or have other malformations such as cleft lip/palate might benefit from interventions designed to counter stigmatization (Morris, Messer, & Gross, 1995) or to enhance social communication (Brinton & Fujuki, 1993).

With respect to subgroups defined by gender and maternal age, case–control differences on the SCT and TRF broadband scales were more pronounced for girls and children of younger mothers, effects that were somewhat more discernible for the TRF internalizing scale than the externalizing scale. There was little evidence for these effect modifications on any parent-report measure. The teacher data are consistent with prior research suggesting that internalizing disorders and symptoms are generally more prevalent among girls with chronic health needs (Ghandour, Kogan, Blumberg, & Perry, 2010). Prior research has also shown that children of younger mothers have elevated risk of poorer outcomes in several domains of functioning including the parent-child relationship (Leadbeater, Bishop, & Raver, 1996), behavioral problems (Fergusson et al., 1993; Wadsworth, Taylor, Osborn, & Butler, 1984) and the child’s neurodevelopmental status and academic achievement (Brooks-Gunn, Guo, & Furstenberg, 1993; Fergusson et al., 1993). It would appear that these well-established correlates of psychosocial adjustment are equally relevant for children with HFM, at least for outcomes reported by teachers. These correlates may create opportunities for early detection and preventative interventions; for example, the poorer outcomes identified for children of younger mothers suggest that interventions targeted to such families could be especially beneficial as previously reported (Speltz, Kapp-Simon, Cunningham, Marsh, & Dawson, 2004).

Limitations

Among the several limitations of this study, families with higher social risk were underrepresented, despite a relatively robust participation rate in relation to the long interval between the original study and follow-up. This affected both case and control groups, but sensitivity analyses (IPW) suggested that group differences remained fairly robust after accounting for differential attrition. Another limitation is the use of parent and teacher questionnaires alone to assess psychosocial functioning; the inclusion of children’s reports would have provided better assessment of symptoms that are difficult for adults to observe (Long et al., 1992). Other methods to assess functioning, such as structured diagnostic interviews, would allow for a flexible, clinician-led approach to better assess psychosocial difficulties in this population (March, Albano, & Riddle, 1996; Stallings, March, & March, 1995). Finally, the use of medical records to garner information about associated conditions and phenotypic characteristics was problematic, given the lack of standardized, detailed reporting across and even within medical centers. Nonetheless, this study
highlights the domains that will benefit from more direct assessment in our future research with this population.

**Conclusion**

Teacher, but not parent, reports suggest that children with HFM have modestly elevated risk for internalizing behavior problems, lower social competency, and less peer acceptance. Children with HFM whose defining features include eye anomalies and those with other malformations such as oral clefts may be at particular risk for developing these teacher-observed problems. Girls and children of younger mothers with HFM may also be more likely to experience difficulties. Further assessment of this cohort is underway to examine neuropsychological, academic, and psychosocial functioning in early adolescence and to determine whether the current measures are predictive of these later outcomes.

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**References**


