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Introduction

Developmental Language Disorder (DLD) refers to the language difficulties experienced by young people that cause barriers to communication or learning in their daily lives, and the language disorder is not associated with a known biomedical condition and: 'where past research has indicated they are unlikely able to make gains spontaneously'.^{1,2} The language difficulties experienced by an individual with DLD involve a deficit in their ability to use and understand spoken or written language,³ and span more than one area of language. These may include: morphology (e.g. structure of words and parts of words), the rules of word and grammatical structure; semantics, rules regarding word meanings and combinations; or pragmatics and, the social use of language.⁴ Children and young people with DLD are at an increased risk of social, emotional and behavioral difficulties and mental health difficulties.⁵⁻⁹ Several recent studies have confirmed that failure to develop adequate language levels in the preschool years is a risk factor for later language difficulties,¹⁰ literacy impairment,¹¹ numeracy impairment,¹² and socio-behavioral aspects of development.¹³ Hence, early identification of the speech and language needs of young people is vital for their health and well-being.

When Language Disorder co-occurs with a biomedical condition such as craniosynostosis, the accepted terminology is Language Disorder associated with Craniosynostosis.^{1,2} An emerging body of literature points to the presence of Language Disorder associated with Craniosynostosis in some children.¹⁴⁻¹⁷ To date, no known research has investigated whether the presentation of Language Disorder associated with Craniosynostosis is distinct from Developmental Language Disorder in the general population.

Many additional factors impact on the communication of children with craniosynostosis, and may be associated with some genetic syndromes, such as oral structural anomalies, hearing impairment, visual impairment, cognitive difficulties.¹⁴ In the United Kingdom, the Craniofacial Specialist Speech and Language Therapists in the four Highly Specialized Craniofacial Units currently undertake regular assessment of the speech and language development of children with craniosynostosis at key developmental milestones. This is in accordance with an agreed, systematic developmental screening protocol to identify the presence of Language Disorder Associated with Craniosynostosis, and other communication difficulties, and permit timely referral to intervention services. The assessment protocol comprises assessments at the time points indicated in Figure 1.

Figure 1. Oxford Craniofacial Unit Speech and Language Therapy Assessment Protocol.

During children's multidisciplinary clinic appointments, parents are also regularly asked by team members about their child's communication development, and whether they have any concerns about their child's speech, language and communication. At the Oxford Craniofacial Unit, formal language assessments are conducted in accordance with a systematic developmental screening protocol

developed by the Speech and Language Therapists in the four United Kingdom (UK) Highly Specialized Craniofacial Centers, and also when clinically-indicated by parental concern, or indicated by screening by the Craniofacial Specialist Speech and Language Therapist. (Figure 2).

Figure 2. Oxford Craniofacial Unit Speech and Language Therapy Support in Multi-Disciplinary Clinics in addition to Protocol Assessments.

The present study aimed to compare parental concern regarding communication development from a binary yes/no question, with the results of a detailed standardized questionnaire about communication, and to investigate whether the parental concern is sufficient to identify the presence of communication difficulties.

Materials and Methods

A retrospective casenote review was undertaken for all patients who attended for a craniofacial multi-disciplinary appointment (June 2017–July 2018). Parents were provided with both the Oxford Craniofacial Unit Pre-Clinic Questionnaire (Figure 1), and the Children's Communication Checklist - 2 (CCC-2),¹⁸ at the time of their multidisciplinary clinic appointment. The pre-clinic questionnaire comprises a short table of pre-determined domains, and a free-text box, for parents to indicate areas of concern that they wish to discuss with the craniofacial multidisciplinary team prior to their appointment. Parental responses to the Oxford Craniofacial Unit Pre-Clinic Questionnaire were recorded in the domains of: hearing, communication development, behavior, physical development, concentration, school and friendships. For the purposes of this study, a parent indicated concern by ticking at least one item on the table in the aforementioned domains.

The areas of concern indicated on the pre-clinic questionnaire were then correlated with the results of a standardized, guided parent questionnaire about children's language development, the CCC-2,¹⁸ to determine which, if any parental concerns were specific and sensitive enough to identify whether patients required further assessment for LDAC. An external researcher not involved in the routine clinical care of patients extracted the data.

Inclusion and Exclusion Criteria

Inclusion criteria were children receiving active craniofacial care via the Oxford Craniofacial Unit for craniosynostosis; who spoke English as their primary language and had complete data on the Oxford Craniofacial Unit Pre-Clinic Questionnaire and CCC-2.

Children with missing pre-clinic questionnaire data or CCC-2 data, or those children who were exposed to a language other than English at home were excluded.

Criteria for further assessment for Language Disorder associated with Craniosynostosis (LDAC)

The criteria for further assessment for LDAC were defined as a General Communication Composite (GCC) standard score 55 or less on the CCC-2 which

indicated a profile suggestive of LDAC, and would warrant further in depth assessment.

Criteria for Parental Concern:

Parental concern was identified by a positive indication in one of the aforementioned domains on the Oxford Craniofacial Unit Pre-Clinic Questionnaire. Concern relating to speech development on the pre-clinic questionnaire was taken to include broad concerns relating to communication development in the absence of any other option to indicate concerns relating to communication on the questionnaire.

Additional qualitative information was obtained from the patient's medical records including: therapy input (e.g. Speech and Language Therapy, Occupational Therapy, etc.), support in school (e.g., Education, Health and Care Plan in place, involvement of Special Education Needs Coordinator; additional communication needs (e.g. Autism Spectrum Disorder (ASD), Attention Deficit Hyperactivity Disorder (ADHD), Hearing Impairment, etc.)

Statistical methods

Descriptive statistics are used to summarize the data (mean, standard deviation for continuous variables, and number (percentage) for categorical. The outcome of interest is a binary variable of language difficulties using a score of 55 or less on the CCC-2 score. Predictors of interest are variables relating to parental concern relating to hearing, speech development, behavior, physical, school work, concentration, friends), informant (mother or father), whether they are receiving local support, additional diagnosis. Logistic regression was used to describe the association of predictors with language difficulties based on the CCC-2 score. Receiver Operator Characteristics (ROC) are used to estimate the sensitivity and specificity of individual predictors with the language difficulties outcomes, and discriminatory ability of predictors through measuring the Area Under the ROC curve (c-statistic).

Results

Participants were parents of 89 monolingual English-speaking children. Sixty-two of the patients were males and 27 females. The patients were in the age range of four to 13 years; mean age of children was 8 years 7 months. The participants were receiving active multi-disciplinary care at the Oxford Craniofacial Unit between June 2017 and July 2018. Eighty-three patients had undergone a transcranial procedure. All patients had a known diagnosis of craniosynostosis – with a variety of sutures affected: sagittal synostosis (clinically non-syndromic) (n=35), metopic synostosis (clinically non-syndromic) (n=17), unicoronal synostosis (clinically non-syndromic) (n=12), multisuture synostosis (clinically non-syndromic) (n=6) and bilateral lambdoid synostosis (clinically non-syndromic) (n=1), Saethre-Chotzen syndrome (n=3), Crouzon syndrome (n=3), Muenke syndrome (n=3), Pfeiffer syndrome (n=1), Turner syndrome (with sagittal synostosis) (n=1), Russell-Silver syndrome (with metopic synostosis) (n=1), Apert syndrome (n=1), Craniofrontonasal syndrome (n=1), *SMAD6* (2 with metopic synostosis, 1 with multisuture synostosis) (n=3), *Z1C1* mutation (with bicoronal synostosis) (n=1) (Table 1).

Oxford Craniofacial Unit Pre-Clinic Questionnaire

Eighty-nine Oxford Craniofacial Unit Pre-clinic Questionnaires were collected prior to children's multidisciplinary clinic appointment from parents (June 2017–July 2018). Of these, 1% (n=1/89) of parents reported they were concerned regarding both speech and hearing development on the pre-clinic questionnaire. Twenty-two percent (n=20/89) of parents indicated concerns about their child's attention and concentration, 22% (n=20/89) were concerned about their child's behavior and 15% (n=13/89) reported concerns about their child's school work.

Results indicated that 78% (n=69/89) of informants were mothers and 21% (n=19/89) were fathers. One informant was a grandparent. One (1%) father indicated concerns about their child's communication development on the pre-clinic questionnaire. Of those fathers who completed the CCC-2, 6/19 (32%) children required additional assessment for Language Disorder associated with Craniosynostosis. 4/69 (6%) mothers reported concerns about their child's communication development on the pre-clinic questionnaire and 22/69 (32%) required additional assessment for Language Disorder associated with Craniosynostosis.

Results from the Oxford Craniofacial Unit Pre-Clinic Questionnaire indicated that 6% (n=5/89) of parents reported concerns about their child's communication development. Results of the CCC-2 indicated that 29/89 (33%) of children had required further assessment for LDAC. A total of 22% (n=20/89) of children were receiving local services and support for a variety of additional learning and communication needs.

Results of Clinical Record Review

Additional qualitative information about participants was obtained from the patient's medical records. All parents reported that their children were attending school in the United Kingdom. Twenty (22%) children received professional and educational support including from Speech and Language Therapy (SLT), additional support for special education needs (SEN), and/or Occupational Therapy (OT).

Clinical notes review indicated that 38% (n=34/89) of patients had craniosynostosis accompanied by an additional communication need such as ASD, hearing impairment, ADHD, Language Disorder Associated with Craniosynostosis, and learning difficulties. Of this patient sub-group, 12% (n=4/34) of parents were concerned about their child's communication development and 18% (n=6/34) of parents reported concerns on hearing on their pre-clinic questionnaire. Sixty-eight percent (n=23/34) of children with additional communication and learning needs were also identified as having communication difficulties on the CCC-2. Of the 23 children with identified communication difficulties on the CCC-2, 47% (n=13/23) were already receiving local support.

Logistic Regression Analysis

Results of logistic regression analysis indicated that parental concern about behavior was the most important factor in identifying whether a child required further assessment for LDAC Odds Ratio (OR) 4.9 95% CI (1.2 to 19.5), $p=0.023$ (Table 2).

Table 2. Logistic Regression Analysis

The strongest predictors of a patient requiring further assessment for LDAC, were if the child was currently receiving additional support OR 5.9 (95% CI 1.5 to 23.9), $p=0.012$, and had an additional diagnosis of Autism Spectrum Disorder OR 15.5 (95% CI 3.1 to 77.4), $p=0.001$ or another diagnosis (e.g. hearing impairment, ADHD, LDAC or learning difficulties) OR 9.5 (95% CI 2.3 to 38.3), $p=0.002$. A regression model containing three variables of parental concern about behavior, access to additional support and the presence of an additional diagnosis, was useful as a diagnostic tool to predict the need for further assessment for LDAC, with good sensitivity (82.8%), specificity (80%) and model discrimination (AUC = 0.88 95% CI (0.80 to 0.96)) (Figure 4). Importantly, parental concern about communication development alone was not sufficiently sensitive or specific to identify patients who required further assessment for LDAC. Results indicated that there were 25 false negatives (i.e. 25 parents identified no concerns about their child's communication development, on the pre clinic questionnaire, and yet further assessment for LDAC was indicated on the CCC-2). Four parents had concerns about their child's communication, which was consistent with identification of requiring further assessment of LDAC on the CCC-2, and one parent indicated concern about their child's communication which was not born out by more detailed assessment on the CCC-2.

Figure 4. Sensitivity and Specificity of Parental Report of Communication Difficulties.

Discussion

We present the first-known study investigating the specificity and sensitivity of parent-reported concern for identifying the need for further assessment of Language Disorder associated with Craniosynostosis.

This study involved a heterogenous group of children with craniosynostosis. The purpose of this study was to better understand the sensitivity and specificity of parental report in identifying children who require further assessment for LDAC in this population. It is of note that our results showed similar findings to previous investigations of language development in children with craniosynostosis. Results indicated that 33% ($n=29/89$) of children presented with communication/language disorder/difficulties on the CCC-2. In our cohort, 17% ($n=6/34$) of children with sagittal synostosis required further assessment for LDAC. Shipster and colleagues identified that 37% of 76 children with non-syndromic sagittal synostosis presented with phonological speech and or receptive and expressive language difficulties.¹⁵ In our cohort, 35% ($n=6/17$) of children with metopic synostosis required further assessment for LDAC. Kini et al. (2010) identified that in 110 children from the Oxford Craniofacial Unit with metopic synostosis, 11 with non-syndromic synostosis presented with 'speech delay' and 10 children with syndromic metopic synostosis presented with 'speech delay'. However, the results of Kini et al. (2010) do not specify whether children's speech sound production or expressive language was delayed.¹⁸ In the present study, 5/6 children with multisuture synostosis presented with LDAC in the absence of any other diagnosis. One of these children has a history of extreme prematurity, the remaining three had a known genetic diagnosis.

In the present study, 33% (n=1/3) of children with Saethre-Chotzen syndrome presented with delayed communication. Kilcoyne et al. (2019) found that 43% (n=13/30) of the children with Saethre-Chotzen syndrome studied presented with receptive and/or expressive language difficulties during childhood.^{13,14} Results of the present research also indicated that 5/6 children with multisuture synostosis, 1/1 children with a left unicoronal synostosis, 1/1 with a right unicoronal synostosis; 1/3 with Crouzon syndrome, 0/3 children with Muenke syndrome, 1/1 with Turner syndrome, 1/1 with Russell Silver syndrome, 1/1 with Apert syndrome, 1/1 with Craniofrontonasal Syndrome, 1/3 with *SMAD6*, 1/1 with bilateral lambdoid synostosis, 0/1 with Pfeiffer syndrome, and 1/1 with *Z1C1* mutation required further assessment for LDAC. All of which have varying rates of reported communication difficulties in the literature.¹⁹⁻²¹ The results of the present study reinforce the importance of regular, longitudinal assessment of children's communication.

Results indicated parental concern about behavior was the best predictor for identifying whether further assessment for LDAC was indicated ($p=0.023$). This is consistent with the literature that reinforces the relationship between language and behavioral difficulties observed in the general population.²² This is of particular importance in children with craniosynostosis where behavior changes are one of the potential signs of raised intracranial pressure.²³ This further consolidates the rationale for regular assessment of children's communication by Craniofacial-Specialist Speech and Language Therapists, to better identify whether behavior changes are related to Language Disorder Associated with Craniosynostosis or symptomatic of possible raised intracranial pressure.

Interestingly, results indicate that parents are aware and concerned about the functional sequelae of language disorder (e.g. behavioral difficulties), but do not necessarily link these concerns with the underlying cause. This is reinforced by the findings that 15% (n=4/26) of parents indicated concerns about their child's communication when the results of their CCC-2 indicated further assessment for LDAC was required. This indicates that 86% (n= 24/28) of children who required additional assessment for LDAC would not have been identified if parental concern was relied upon alone. The implications for clinical practice are clear: relying on parental report of concern about communication is insufficient to identify the presence of Language Disorder Associated with Craniosynostosis, necessitating regular assessments by Craniofacial Specialist Speech and Language Therapists.

What is surprising is how few parents of children with additional diagnoses, and associated communication difficulties, such as ASD and ADHD identified from their child's medical notes were concerned about their child's communication (4/34 parents indicated concern). This could be because their children were already receiving adequate support (n=15/34). However, fifteen children with parents who did not indicate concern about their communication, had an additional diagnosis and were not currently receiving support. Literature on The International Classification of Functioning, Disability and Health (ICF), 2001 by the World Health Organisation revealed parental perceptions and concerns of children with multiple learning difficulties are more related to the body impairments of their children when compared to communicative skills/activity limitations.²⁶ However, studies have found parents to be reliable in their observation of their child's overall development.^{24,26-28} These studies also reveal parental participation in the assessment process promotes a

collaborative initiative of professionals working closely with parents to detect their child's communication needs. This partnership is therefore a great source of support that the multidisciplinary team can harness in the decision-making process.

Existing information regarding the specificity and sensitivity of parent's perceptions of the presence of DLD in children in the general population is limited. Research has largely focused on the use of guided, parental questionnaires to identify DLD in children.²⁹ The only known literature relating to parental concern and communication development has investigated parent-perceived hearing loss, and highlighted that parents were unable to reliably report hearing loss in their child.^{28,30,31,32} This is the first known study to investigate the specificity and sensitivity of parent's perceptions of the presence of Language Disorder associated with Craniosynostosis.

It is generally agreed that no single measure is sufficient to capture the multi-dimensional nature of language.²⁹ There are three general approaches to the assessment of language in children: standardized norm referenced assessments, analysis of language samples, and parent/carer reports of language performance or concern about language skills.²⁹ In this study, we have investigated whether initial parental concern related to communication development asking a binary question (Yes/No), is sufficient to identify the presence of communication difficulties, compared to completion of a detailed, standardized questionnaire about communication. Results indicate that concerns about behavior may indicate the need for further, detailed language assessment. Whilst difficulties with behavior and language are not mutually-exclusive, the established link between the two domains necessitates careful monitoring. In many children, behavior is a form of communication when they are otherwise unable to express themselves.

A limitation of this study is that the question on the pre clinic questionnaire relates to speech concerns, and it is not known if parents would have endorsed this if they had concerns about children's social use of language. The pre-clinic questionnaire has now been revised to permit parents to indicate if they have concerns about a child's understanding and use of language. **All clinicians in the Oxford Craniofacial Unit multi-disciplinary team have been made aware of the sensitivity and specificity of parental concern about behavior for identifying whether further assessment for LDAC is required, and are able to refer to the Speech and Language Therapy team if concerns are raised outside of protocol ages. The sample size of the present study precluded further statistical analysis by craniosynostosis diagnosis. A direction for future research would be to conduct sub-group analysis to determine whether the sensitivity and specificity of parental concern differs according to craniosynostosis diagnosis.**

This paper provides clinicians in craniofacial centers without access to Speech and Language Therapy support with important knowledge about when a referral to Speech and Language Therapy in alternative sectors (such as education) may be warranted. In summary, if parents of children with craniosynostosis indicate concerns about their child's behavior, this may indicate that the child has LDAC, and a referral for further assessment by Speech and Language Therapists should be considered.

Conclusion

The results of this study show that the need for further detailed assessment for Language Disorder Associated with Craniosynostosis was indicated through parental concern about behavior, therefore wherever concerns about behavior are raised; clinicians should consider whether Language Disorder associated with Craniosynostosis is an underlying factor. Results also indicate that parental concern alone is not sufficient to identify language disorder, and that regular formal assessment is required. The link between behavior and language previously reported in the literature is reinforced.

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Figure 1. Oxford Craniofacial Unit Speech and Language Therapy Assessment Protocol.

Figure 2. Oxford Craniofacial Unit Speech and Language Therapy Support in Multi-Disciplinary Clinics in addition to Protocol Assessments.

Figure 3. Oxford Craniofacial Unit Pre-Clinic Questionnaire

Figure 4. Sensitivity and Specificity of Parental Report of Communication Difficulties.

Table 1. Patient Characteristics, Domains of Concern on the Oxford Craniofacial Clinic Pre-Clinic Questionnaire, and General Communication Composite score on CCC-2.

Table 2. Logistic Regression Analysis