

# Communication

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## Introduction

Audrey Thurm, PhD presented on communication in individuals with PMS, in a talk entitled “Let’s Talk about More than Just Talking: Language, Communication, and Social Behavior in PMS”. This presentation provided an overview of important definitions as well as current literature on language, communication, and social communication in PMS.

Language is a method of communication, which may either be in written or spoken form, which uses words in a structured and conventional way. Receptive language refers to one’s ability to understand other’s use of language, whereas expressive language refers to one’s ability to produce language. Communication refers to the exchange of ideas through verbal and/nonverbal means. Social communication is a specific form of communication that is used in social situations, that typically involves reciprocity and the sharing of information between at least two individuals.

Among individuals with PMS, language skills are highly variable. In terms of expressive language, data from prior literature as well as the PMS Registry indicate that approximately 75% of individuals with PMS are considered to be “minimally verbal”, which is defined as little expressive spoken language abilities by school-age. This is in contrast to the approximately 25% of individuals with ASD who are considered to be minimally verbal. With regard to receptive language, individuals with PMS demonstrate significant variability, with some individuals showing difficulty responding to their name and simple one-step instructions and others being able to follow multi-step instructions. There are findings suggesting that language abilities may be related to deletion size; however, results remain disputable. More consistent findings demonstrate a relationship between language abilities and comorbidities of developmental, intellectual, and autism spectrum disorders (i.e., greater language impairment related to presence of comorbidities). Additionally, available data suggests a potential relationship between motor and language abilities in individuals with PMS, such that worse motor abilities may be predictive of more severe language deficits.

Communication is a more difficult skill to assess in individuals with PMS, firstly because it is a relatively broad concept and secondly because standardized measures of communication are limited. Thus, communication is typically assessed in individuals with PMS via caregiver report (e.g., Vineland Adaptive Behavior Scales, Autism Diagnostic Interview-Revised) or direct observation. These assessments may examine early communicative behaviors (i.e., body movements, babbling, facial expressions, visual gaze, and simple gestures) as well as higher-level communicative behaviors (i.e., conventional gestures, concrete and abstract symbols, and spoken language).

Research clinicians from University of Kansas have developed research using the Communication Complexity Scale (CCS) to measure the potential of communicative behaviors. This tool assesses the presence of early communicative behaviors, such as orienting, scanning, and triadic gaze, in order to predict future communication abilities. For example, Crais and colleagues (2004) reported that communicative behaviors demonstrated on the CCS related to development of typical gestures later in life.

Social communication is similar to communication with the primary exception that the communication strategies are specifically used within a social context. An example of lower-order social communication skill is requesting and an example of a higher-order social communication skill is initiating joint attention. Presently, social communication skills are assessed using behavioral and/or parent-based methods, including the Autism Diagnostic Observation Scale and the Autism Diagnostic Interview-Revised, respectively. Yet, these tools have their own limitations in their utility in the PMS population as it is not well-known how the severity of intellectual disability may impact core social communication symptoms associated with ASD. Thus, the development of adapted measures more appropriate for the PMS population (i.e., minimally verbal) is advised.

Finally, due to limitations of spoken language and other forms of communication for the majority of individuals with PMS, many families elect for their children to use alternative or augmentative communication devices. Common methods include Picture Exchange Communication System (PECS) and electronic devices such as Dynavox and application-based programs (e.g., Proloquo). Some families also choose sign language. Despite the wide variety of alternative communication methods, there is a limited number of qualified individuals to provide individualized and sustained training on use of these methods to individuals with PMS and their families.

## Identified Problems

- 1. My child does not speak or is minimally verbal. I do not know which alternative communication methods/devices that my child should use, and I do not how to use them.**

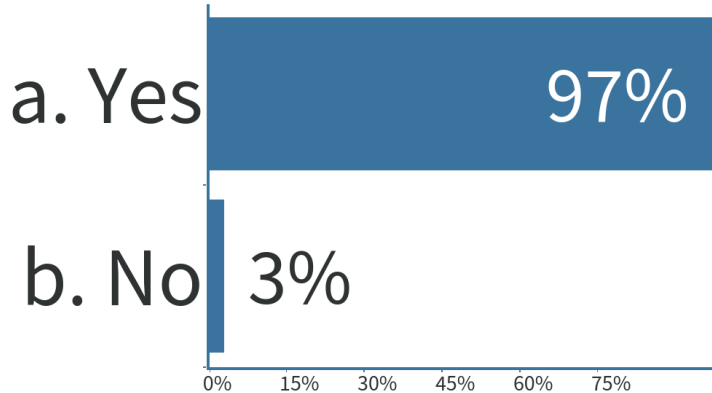
Dr. Thurm reported that based upon PMS Registry data, approximately 75% of individuals with PMS are considered to be “minimally verbal”. This suggests that these individuals need alternative methods to communicate their needs and experiences with others. According to the PollEverywhere poll, 97% of parents indicated that their child communicated through non-speech methods (see Question 1 graph below). However, as observed in the pie-chart figure, the type of alternative communication used by individuals with ASD varies substantially (see Question 2 graph below). Six methods of alternative communication were most commonly identified: traditional Picture Exchange Communication System (PECS), electronic- or app-based PECS, sign language, modeling techniques, repetition techniques, or specific methods taught within

ABA/speech therapies. Of these methods, parents identified ABA/Speech therapies as being most helpful (20%) followed by repetition-based techniques (15%) and electronic- or app-based PECS (13%). However, if you combine PECS categories, this method becomes the most effective method of communication for individuals with PMS (21%) according to parent report. These results should be interpreted with caution, as it is not known exactly which techniques are used within ABA/Speech therapies (i.e., PECS, sign language), and “repetition” is vague. It should also be noted that 6% of parents indicated other methods that did not fall into one of these more specific categories. For example, one parent shared that “nontraditional cues from your child” was most effective for his/her child. Also, 3% of parents indicated a combination of methods was most effective. For instance, one parent reported using both PECS and modeling techniques. Lastly, 20% of parents reported being “not sure” which method was most effective with their children. For example, one family expressed, “I don't know and we struggle with convincing [our child's speech/language therapist] to work on functional communication”. Thus, it appears that even though the majority of individuals with PMS used alternative means to communicate, no single or combination of method(s) is most effective or consistently used across families. Additionally, as the last quote indicated, parents have difficulties working with therapists to identify potential alternative methods of communication. As one parent expressed, “[I am] concerned that the schools and therapists are not willing to try different methods that might be useful, [which] limits [my] child's development”. Furthermore, during the Group discussion, parents also shared the challenges related to teaching their children alternative methods (72 parents indicated this as a concern, 4.6 out of 5) as well as learning these alternative methods themselves (38 parents indicated that this was a concern, 5 out of 5). For example, several parents indicated that their children use app-based PECS, but they themselves are not familiar with the program well enough to use effectively in the home environment. An informal poll completed at one of the roundtables indicated that the degree of formal training that parents received on their children's alternative communication methods varied substantially from none to several hours a month in a structured environment. It was noted that the quality of training variably substantially as well. As one roundtable summed up their needs within this domain, “Families want to be informed about all methods of social communication so they can trial and error before deciding on a method; finding schools/therapists have set ideas for specific communication methods without exploring options”.

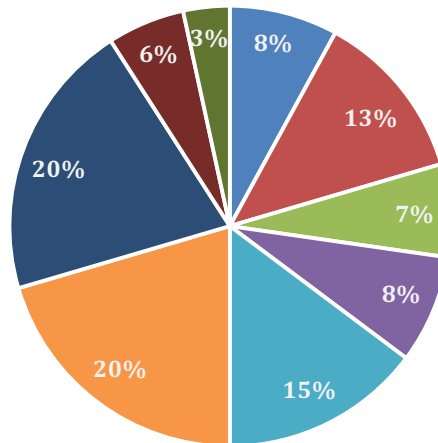
**1) Does your child communicate his/her needs in ways other than speech**

When poll is active, respond at [PollEv.com/pms](https://www.poll-ev.com/pms)

Text PMS to 22333 once to join



**2) What are thought to be the best methods for teaching communication in children with Phelan-McDermid Syndrome?**



- Traditional PECS
- Electronic-/App-Based PECS
- Sign Language
- Modeling
- Repetition
- Not sure
- ABA/Speech Therapy
- Other
- Combination

**2. My child experienced a regression in his/her language skills, and I am lost on what to do next OR I know regression occurs in individuals with PMS, but how do I predict/prevent this in my child.**

Concerns related to regression have been indicated repeatedly throughout the McPosium; however, discussion of these concerns have been restricted to the Social

Communication and Natural History sections as these were the areas with the highest percentage of parent concerns. Thirty-two parents indicated that this was a concern and ranked it to be a significant concern (4.6 out of 5). One specific concern related to regression introduced by several parents was whether alternative communication devices could "cause" regression of children's development of verbal language skills. Similarly, other parents indicated that their children's therapists indicated that only one form of communication (i.e., just speech OR just PECS or just sign language) should be taught for a similar reason. Lastly, several families indicated wanting to know how to predict whether their children would experience regression or not and how to prevent it from happening.

### **3. My child's motor issues are impacting his/her ability to communicate.**

This was an area of concern raised by a smaller proportion of parents (n = 21); however, it was ranked as a significant concern (4.6 out of 5). This suggests that although this may be less common issue for individuals with PMS or concern for parents of individuals with PMS, it is a highly significant issue when it is present. One parent shared these challenges that appears to capture this issue well, "My child is severely delayed in gross and fine motor [skills] and is nonverbal. We've tried several communication devices but no luck. Sign language is not feasible due to lack of fine motor and cognition is very low. [This has a] huge impact because [our child] cannot communicate properly. [It's a] guessing game." Other parents expressed their frustrations related to their children's therapists paying "little attention" to their motor skills, which may impact ability to communication either verbally or through alternative communication devices. Lastly, several parents questioned whether technology used to track eyes as alternative to pointing would be an appropriate, feasible, and effective means to aid in their children's communication.

## **Proposed Solutions**

### **1. Development of practice parameters for Social Communication.**

Due the variability of communication strategies used by individuals with PMS with variable success and quality of implementation, the development of practice parameters on this topic would be of great service for individuals with PMS and their families. Several families during the Group discussion specifically identified the need for practice parameters in this area, and also indicated that parents of older individuals with PMS should be involved in its development. In order for these practice parameters to be most meaningful, additional research should be conducted to assess the effectiveness of the above-mentioned methods. The practice parameters should include not only types of therapies/methods that are effective in this population, but basic characteristics of each to ensure quality control. For instance, specifications regarding minimum training hours for the therapist implementing the therapy/method/device, the child receiving the therapy/ method/device, and the parent using the therapy/method/device at home. The practice parameters also should detail which therapies/method/device would be most

helpful for a particular child based upon the child's characteristics (e.g., prior vocalizations versus no vocalizations; no motor delays versus minor motor delays versus significant motor delays). This may have parents choose which therapy/method/device to try first based upon which is likely to have the most success. Additionally, practice parameters should clarify issues related to regression, using multiple therapies, and bilingualism.

## **2. Increased research into language regression in PMS.**

Due to the overwhelming concern and/or experience of language regression in individuals with PMS, this is an area where additional research is warranted. Specific research aims should include, but not be limited to: identifying time course of regression, identifying risks for regression, determining pattern of recovery, and identifying most effective means to return to baseline. Additionally, increased awareness and support for parents who have/may experience language regression should be a goal as this experience can be frustrating and frightening for parents.

## **3. Increased development and implementation of alternative communication methods for individuals with severe motor limitations.**

Language delays and difficulties may be further exacerbated by motor delays and difficulties; thus, it is necessary to develop and implement alternative communication strategies that can be used by individuals with PMS who are unable to use more traditional means of communication/alternative communication. The use of such devices have successfully be implemented in individuals with disorders that severely impact motor functioning (e.g., cerebral palsy). Thus, it may be important to examine the literature and practices in populations outside PMS and ASD. As methods are identified or developed, the use of these methods should be disseminated to the PMS community and their implementation should be promoted to both families and providers (e.g., therapists, teachers). Furthermore, greater awareness of the impact of motor disabilities on communication also should be a goal. As part of this goal, clinicians who are assessing and treating individuals with PMS should specifically assess for motor delays/abnormalities, and determine how these motor differences may impact communication functioning. Providers then should work with families to identify whether communication methods need to be appropriately adapted with these motor differences in mind. It also may be important to incorporate or add motor therapies (i.e., occupational and physical therapy) to a child's intervention programming within an outpatient and/or in-school setting.