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## **Minding the Gap: A Large Scale Survey of Agenesis of the Corpus Callosum and Other Callosal Anomalies**

**By Gary Schilmoeller & Kathy Schilmoeller**

*Thank you to all who conscientiously filled out this long survey. We also appreciate all the family members, friends, and professionals who share their love and expertise with those with ACC. And we thank each of the children and adults with ACC for being our teachers.*

*We thank Warren Brown and Lynn Paul of the Travis Institute for Biopsychosocial Research, Pasadena, CA and Paul Moes of Calvin College, Grand Rapids, MI for their valuable help in the development of the survey. Finally, we thank the University of Maine for supporting this project with a Faculty Research Fund grant.*

### **Introduction**

Existing information about agenesis of the corpus callosum (ACC) comes mostly from the medical field and the studies usually are based on very small numbers of participants (e.g., O'Brien, 1994; Wisniewski & Jeret, 1994). Noting that some professionals suggest that people who have only ACC may be asymptomatic - that is, they will develop normally and without delays, O'Brien argues that more comprehensive studies need to be conducted in order to test this hypothesis. He conducted a study of 47 children with ACC with the intent of beginning to describe the behavioral and developmental consequences of ACC (O'Brien, 1994). Our intent in this study was to expand upon O'Brien's work with a more comprehensive survey of a much larger group of people with ACC.

### **Sample**

In March, 2000, we mailed a comprehensive survey to 1,900 families who had a member with ACC. Six hundred and seventy eight (36%) of the surveys were returned. Most of the respondents were birth mothers (89.2%) with a small number of birth fathers (4.9%) and mothers of adopted children (3.0%). The mean age of the mothers of the person with ACC was 37 years old; the youngest mother was 19 and the oldest was 68. The mean age of the fathers was 40; the youngest father was 20 and the oldest was 75. Respondents had completed an mean of 14.9 years of school (ranging from completing ninth grade through more than 18 years of education). Their spouses had completed a mean of 14.7 years of school (ranging from completing sixth grade through more than 18 years of education).

The mean age of the persons with ACC was 7.6 years (ranging from 4 months to 45.5 years). Nearly fifty-eight percent (57.9%) were males and 42.1% were females. Most of the persons with ACC (91.3%) were white. A small number were Hispanic (3.9%) and Asian (2.0%). The rest were either Black, Native American, or listed their race or ethnicity as some other category.

## Survey

The survey was constructed based on descriptive information published in earlier survey studies of people with ACC (e.g., O'Brien, 1994; Schilmoeller & Schilmoeller, 2000; Schilmoeller, Schilmoeller, & Baranowski, 1999) as well as from reports of parents who had shared information about their child with ACC through phone calls, emails, and regular mail correspondence with us at The ACC Network or through themes that appeared in discussions on the ACC-L listserv, an electronic discussion group focused on issues pertaining to ACC. Categories of questions focused on general diagnostic information as well as the physical, social, and communication skills of the persons with ACC. Types of learning style, settings where these children received education, and therapies received also were described.

## Results

Diagnostic History. The most frequent method of diagnosis was magnetic resonance imaging [MRI] (82.7%). Fifty percent of the parents reported that a CT scan was the method diagnosis. Ultrasound (26.9%) and EEG (21.0%) were also used to diagnose the ACC. The percentages add up to more than 100% because many children had more than one method used to determine and then corroborate the diagnosis.

Concurrent Conditions. The ten most frequently reported concurrent conditions were developmental delays (78.0%), learning disability (32.8%), seizures (29.9%), mental retardation (26.8%), cerebral palsy (16.1%), microcephaly (14.1%), hydrocephaly (12.8%), autism or autistic-like behaviors (9.9%), obsessive compulsive disorder (7.0%), and attention deficit hyperactivity disorder (6.7%).

Physical characteristics. Nearly half (46.7%) of the children were described as having a body size that was small or slightly smaller than average compared to others of the same age and gender. On the other hand, the number of children who were described as having smaller than average head size was nearly equal to the number described as having larger than average head size (31.6% v. 30.8%) with the remaining 37.6% described as being comparable in head size to others of the same age and gender. Asked about head and facial anomalies, 17.8% reported the person with ACC had eyes set wide apart, 19.6% had low-set ears, and 23.3% had unusual head shape. The incidence of cleft lip or cleft palate was very low (0.7% and 4.3% respectively). Over half of the respondents (53.9%) reported that their children were either slightly floppy or very floppy (hypotonic) in terms of muscle tone.

One physical characteristic often reported anecdotally by parents is that their children have a high tolerance for pain. For example, parents have reported a child putting a hand on a hot stove burner and not withdrawing the hand nor showing any sign of registering pain, a child getting a hand caught in a car door without protest, and a child with a broken collarbone that was not detected until several days after the injury. While this characteristic is not reported in the research literature about ACC, the anecdotal reports occur frequently enough that we included several questions about sensitivity to pain, touch, and cold. Respondents reported that over half (56.1%) of the children showed either little or no pain perception (19.8%) or less pain perception than average (36.3%). On the other hand, over 40% of the children were more sensitive to being touched by others (30.5%) or much more sensitive to being touched by others (10.0%).

There was no discernible pattern of children being either more or less sensitive to cold than average.

Developmental milestones. Since 78% of the children experienced developmental delays, we asked whether or not children had met several milestones and at what age.

<u>Milestone</u>	<u>Percentage Met</u>	<u>Mean Age Met</u> <u>In Months</u>
Lifted head	62%	5.6
Rolled over	67%	9.9
Sit alone	66%	12.3
Crawled	56%	19.2
Stood alone	47%	21.7
Walked alone	55%	25.3
Talked single words	42%	23.4
Toilet trained	34%	49.9
Rode a bicycle	13%	84.9

Eating and Elimination problems. Parents reported their children had multiple problems with eating. For example, 55% had an inadequate sucking reflex at birth, and 16% had or have a gastric feeding tube. Parents also reported that their children either “occasionally,” “often,” or “in the past” had difficulty swallowing (41.3%), experienced reflux (43.9%), did not know when the child had enough food or fluid (36.9%), tried to eat things other than food (24.9%), or had chewing difficulty (51.9%). Many children had trouble with bladder control (57.2%), constipation (61.4%), or diarrhea (38.8%).

Social Behavior. Parents rated their children’s social behavior on a five point scale - much less, slightly less, about the same, slightly more, much more than others. These children with ACC were slightly or much more likely to enjoy social interaction (44.0%) and be friendly with strangers (42.5%). They also were less likely to be reserved or shy (47.5%) than others. On the other hand, only 21.8% were more likely to enjoy interacting with a peer group. This corroborates parental reports that their children often get along well with adults and younger children, but not as well with age peers. Forty percent of the children also had difficulty using appropriate physical space. This corresponds to the 40% who were more sensitive to touch by others, raising the question of whether sensitivity to touch might play a role in the less frequent social interaction with peers.

Parents rated disruptive behavior on a three point scale - rare/not true, occasional, or very frequent. Some parents reported their children occasionally or frequently physically attacked other people (21.5%), suddenly lashed out for no apparent reason (25.4%), or deliberately destroyed things (16.6%).

Children were rated as often or almost always happy and cheerful (91.0%), content (87.0%), and relaxed (72.5%). By contrast, only 6.4% were sad or unhappy and only 10.6% were angry. Some children showed fearfulness or anxiousness (17.4%), experienced quick mood shifts (22.0%), or showed moods out of place (11.7%).

In summary, children with ACC tended to be rated as very happy, socially-engaging children though they tended to be less social with age peers and to occasionally have problems with anger control.

Communication Skills. Parents reported that the vast majority of children with ACC (81.6%) showed at least some ability to understand from communication, ranging from understanding single words to understanding most messages that are at the person's age level. Indeed, 60.3% were able to understand either most long sentences or understand most age-level messages. In terms of expressive communication, though, 33.9% were not able to communicate verbally at all. And, in contrast to the 60.3% who could understand long sentences or age-level messages, only 40.9% could express such verbally.

Communications by these children were sometimes challenging. For example, children occasionally or frequently engaged in meaningless conversation (38.1%), shouted or screamed unexpectedly (39.9%), engaged in out-of-place conversations (45.2%), or repeated words or phrases apparently without understanding them (30.5%).

Learning Variables. According to their parents, children with ACC "often" or "almost always" learn by repetition (84.1%) and by imitation (66.3%). They also have good memories (58.2%). And many enjoy learning and working on computers (57.5%).

On the other hand, these children have difficulty with abstract reasoning (67.6%), have difficulty staying on task when learning (54.0%), and persevere on some details when learning something (42.0%).

Parents report a variety of settings in which learning can occur for their children with ACC including play groups (40.1%), preschools (56.3%), self-contained special education classrooms (46.1%), regular education classrooms (32.4%), resource room (sometimes called "pull-out" programs) (19.4%), and other types of special education programs (20.0%). A small percentage of children attend Headstart programs (8.0%) or are home-schooled (7.9%). Often a single child experiences a variety of these programs, sometimes at the same time and at other times sequentially as the child gets older. Nearly one-fifth (18.3%) of the children were too young to begin any form of schooling.

Therapies and Intervention Services. More than half of the children with ACC received three therapies - speech therapy (65.1%), occupational therapy (64.1%), and physical therapy (60.5%). Other therapies commonly reported were early intervention (36.9%), sensori-integration (36.3%), academic counseling (23.9%), and vision therapy (18.0%). Early intervention also was reported to have been received "only in the past" by 45.1% of the families. Thus, speech therapy, occupational therapy, physical therapy, and early intervention are the therapies most frequently provided for these children.

### **Summary Comments**

These data represent only a very preliminary analysis. In subsequent analyses, we will look at these variables for different age groups of the children with ACC. We will compare those with complete ACC and those with partial ACC to see whether there will be differences between groups on some of the categories we reported here. We also will try to distinguish between children who have ACC and many other concurrent conditions with those who have only ACC and no other or only a few other concurrent conditions. Results of these and other subsequent analyses will be published in later newsletters.

## References

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## Working with Your Speech-language Pathologist to Evaluate the Communication Skills of the Child with ACC

By Judith Stickles, M.A., CCC-SLP

In eighteen years of working as a speech-language pathologist (SLP), I have had the opportunity to work directly with a young man with agenesis of the corpus callosum (ACC) over a twelve-year period. Through my association with the ACC Network, I also have had the opportunity to consult with many other speech-language pathologists concerning individuals with ACC. I have been impressed with the wide range of communication issues that arise with individuals with ACC. Whether your child is nonverbal or displays subtle pragmatic or social problems, it is important for a speech-language pathologist to assess the child's communication skills as early in the education process as possible.

The evaluation process at its best is collaborative. Here are some suggestions of what a parent/ caregiver can contribute to the evaluation:

- Bring a list of concerns. Write down all of your concerns and questions so the SLP can directly address them orally or in the evaluation report.
- Describe your child's medical condition. Bring or mail ahead all relevant medical reports. Many SLPs are not familiar with the condition of ACC, so a brief summary of what it is and how it affects your child would be helpful. A copy of a CT scan or MRI and neurological report would also be useful.
- Describe early experiences concerning feeding. Since oral structures involved with speech production and

feeding are similar, information regarding early feeding experiences is important. Was your child able to breastfeed or suck from a bottle with ease? When did your child manage solid food and feeding independently? Was feeding a pleasant experience for you and your baby? How does your child eat now? Are there any foods or liquids that are avoided? Does your child ever choke or cough when eating?

· Describe early communication experiences. Developmental milestones such as when the first word and word combinations were spoken are important as well as descriptions of early play styles and preferences and interactions with siblings and others. What were some of your child's first words and word approximations (for example, sounds your child made consistently to represent an object like "ba" for ball)? Did he or she have preferences for certain words and sounds? When your child acquired new words, did they remain in his or her vocabulary or disappear not to be heard again? If your child is not speaking, focus on describing how he or she communicates nonverbally such as by eye contact, body motion, and gesture. Any videotapes of your child at play or audiotapes of your child talking would be very useful.

· Describe a typical day at home or school for your child. What kinds of communicative opportunities exist? With whom does your child appear most comfortable communicating? Does he or she have opportunities to communicate in formal contexts such as a classroom or church in addition to the informal context of home?

The nature of the speech-language or communication evaluation will depend on the particular needs of your child. For the child who is

nonverbal, the SLP will help to determine why the child is not speaking and help you develop an effective communication system with your child, building upon what is already working for you both. The Hanen Early Language program {252 Bloor Street West, Suite 3-390, Toronto, Ontario, Canada M5S 1V5, (416) 921-1073} is a wonderful resource for SLPs and parents to use with preschool children who are not yet verbal or are struggling with developing early speech and language skills. It focuses on language stimulation techniques that can be used by families in everyday contexts at home and preschool.

If your child is nonverbal or significantly delayed in acquiring verbal skills, your SLP may encourage the use of another form of communication such as sign language or a picture board. Usually these forms of nonverbal communication are used in addition to other language stimulation techniques to augment the child's communication system. Often their use decreases communicative frustration and in some cases actually stimulates oral language development.

My review of several case studies involving individuals with ACC indicates that your child with ACC is "at risk" for certain specific communication problems. These areas include developmental apraxia of speech, delays in the acquisition of phonological skills, and difficulties in the acquisition of pragmatic or social communication skill.

### **Developmental Apraxia of Speech**

This is a speech disorder that occurs because there is a break-down in the motor programming for speech. The child is hearing and perceiving sounds accurately but cannot make the

articulators consistently produce the sounds. Some clues that your child may have developmental apraxia of speech include generally poor speech intelligibility (you can understand very little of what your child says), many moments of pausing when he or she is talking and groping behaviors with the mouth when words are attempted, an overall monotone quality to speech production, and an overall inconsistency with speech errors (instead of always saying w/ r as in wed/ red she produces the word a different way with each attempt). Developmental apraxia of speech is a serious communication problem involving both speech and language; however, most children respond to frequent, consistent speech therapy which should begin as early as possible.

### **Phonological Awareness**

Phonological awareness refers to the conscious awareness of the sound structure of language including knowledge that words are composed of syllables and sounds. In the past two decades, research in many educational fields has supported a strong correlation between phonological awareness skills and reading success. The acquisition of phonological awareness skills begins in the preschool years and continues as the child progresses through school. Many schools now include a screening of phonological awareness skills as part of the kindergarten screening process. Since your child with ACC may be “at risk” for difficulties in this area, a speech-language evaluation should include a test of these skills. If your child is having difficulty with reading, writing, and/ or spelling, an evaluation of phonological awareness skills should occur. Appropriate tests of phonological

awareness include The Phonological Awareness Test (Robertson and Salter), Lindamood Auditory Conceptualization Test (Lindamood and Lindamood), or The comprehensive Test of Phonological Process in Reading (Wagner and Torgensen).

### **Pragmatics/Social Skills**

In reviewing case studies of children and young adults with ACC, the most commonly reported communication difficulty was in the area of pragmatics/ social skills. These problems may be apparent in the toddler/preschool years or may not surface until the language of socialization becomes more abstract and complex in the early adolescent years. If your child is experiencing difficulty with peer relationships, it is important to have a social skills assessment by an SLP. There are very few standardized tests that examine pragmatic skills. Some that may be helpful are The Test of Pragmatic Languages (Phelps-Teraski and Phelps-Gunn) and The Test of Problem Solving (Zachman, Huisinigh, Barrett, Orman, and LoGiudice). It is important to remember, however that these tests create artificial contexts for language, and the child should be observed in real social situations with their peers as well. I recently had a child complete with great accuracy all items on the Test of Pragmatic Language. When I complimented him on his performance, he responded by saying “I know those are the things you’re supposed to say, but those aren’t the things I say with my friends!” In this case, it was very important also to observe him playing and conversing with friends and to take samples of his language in these contexts.

Standardized tests are important instruments to use in assessing language skills and are required in most school districts for Special Education identification purposes; however, they have their limitations, especially when examining social skills. SLPs can also use what are considered informal language samples, in which the SLP records what the child is saying in at least two contexts (for example, conversing with friends in the classroom or conversing with a parent) and a narrative language samples in which the clinician records the child re-telling or creating a story or giving an explanation. These samples are then transcribed and analyzed by the SLP. Both of these measures examine the child's language skills at the discourse level as opposed to the word or sentence level as with most standardized tests. Languages sampling in particular also provides a sample of language from everyday contexts and may give a more accurate pictures of your child's language skills. Language sampling and narrative sampling are time consuming and work intensive for the SLP but provide valuable information regarding your child's expressive language skills and are measures that should be included in a comprehensive speech-language evaluation.

Finally, it is important to share with your SLP your knowledge of your child's condition and your observations of his or her communication skills as well as the fact that given the diagnoses of ACC your child may be at risk for developmental apraxia of speech, difficulties with phonological awareness skills and/ or pragmatic skills. Early identification and treatment of ay communication difficulty is critical to academic and social success. The

process is enhanced when parents are active participants on their child's education team.

**If you need information to help you locate a qualified SLP in your area, contact the following resources:**

Argentina – Asociacion Argentina de Logopedia, Foniatria y Audiologia, phone: 4815-5997, email: [asalfa@ciudad.com.ar](mailto:asalfa@ciudad.com.ar)

Australia – Speech Pathology Australia, phone: +61 3 9642 4899, email : [office@speechpathologyaustralia.org.au](mailto:office@speechpathologyaustralia.org.au)

Canada – Canadian-Speech-Language-Hearing Association, phone: 800-259-8519, url: [www.caslpa.ca](http://www.caslpa.ca)

Hong Kong – Hong Kong Association of Speech Therapists, email: [hkasts@netvigator.com](mailto:hkasts@netvigator.com)

Japan – Japanese Association of Speech-Language-Hearing Therapists, email: [ZAJOO@nifty.ne.jp](mailto:ZAJOO@nifty.ne.jp)

Korea – Korea Speech and Hearing Association, phone: +82-53-629-7322, email: [dhkwon@biho.taegu.ac.kr](mailto:dhkwon@biho.taegu.ac.kr)

New Zealand – New Zealand Speech-Language Therapists Associate, phone: +64 3 235-8257, email: [exec@nzsta-speech.org.nz](mailto:exec@nzsta-speech.org.nz)

Saudi Arabia – Jeddah Institute for Speech and Hearing, phone: (966-2) 667-5311, email: [info@jish.com](mailto:info@jish.com)

Singapore – Singapore's Speech, Language and Hearing Association, phone: 3214549, email: [speech@pacific.net.sg](mailto:speech@pacific.net.sg)

South Africa – The South African Speech-Language-Hearing Association, phone: (011) 726 5014, url: [www.saslha.org.za/index.htm](http://www.saslha.org.za/index.htm)

Taiwan – Professional Association of  
Speech-Language Pathologists in  
Taiwan, email: [shchen@vghtpe.gov.tw](mailto:shchen@vghtpe.gov.tw)

United Kingdom – United  
Kingdom's Royal College of Speech and  
Language Therapists, Switchboard in the  
UK: 020-7378 1200, email:  
[postmaster@rcit.org](mailto:postmaster@rcit.org)

United States – American Speech-  
Language-Hearing Association,  
phone: 301-897-5700, url:  
[www.asha.org](http://www.asha.org)

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