Comparison of Assessment Results of Children with Low Incidence Disabilities

Dennis J. Campbell
University of South Alabama

AmySue Reilly
Auburn University

Joan Henley
Arkansas State University

Abstract: This paper describes a research study that assessed young children with a low incidence disability, specifically Cri-du-Chat Syndrome (CDSC). A description of the concerns of assessing individuals with low incidence disabilities is described. Parent reports (using the Development Observation Checklist System) on the functioning of their children are analyzed and compared to data collected from more formal assessment (specifically, the Battelle Developmental Inventory). Data collected from parent report and formal assessments are included. A result of the analysis shows that parents’ reports are congruent with formal assessment data.

The Individuals with Disabilities Improvement Act (IDEA, 2004) defines low incidence disabilities as “a visual or hearing impairment, or simultaneous visual and hearing impairments; a significant cognitive impairment; or any impairment for which a small number of personnel with highly specialized skills and knowledge are needed in order for children with that impairment to receive early intervention services or a free appropriate public education” (20 U.S.C. 1462 § 662(c)(3)). Since 2002, the No Child Left Behind (NCLB) legislation mandates that all students, including those with disabilities, are educated in the framework of high standards and related to level of performance similar to their general education peers. These two major pieces of legislation are directly impacting the assessment practices for students with low incidence or more severe disabilities and their families.

Many states have developed and implemented a means of assessing students with more severe disabilities by using various alternative means of evaluation procedures (Elliott & Thurlow, 2006). These alternative means of assessment are for those students who are designated by the federal regulations as qualified to be reported by the states as not able to complete the district assessment even with accommodations. Most children served through early childhood special education programs are labeled developmentally delayed (DD), which in and of itself qualifies for an alternative assessment. Many states have elected to use the DD label for children from birth to age five, while other states have opted to use DD for children from birth to age nine. This variance in the use of the DD label impacts the overall reporting by states on their annual yearly progress of assessment options.

IDEA 2004 states that a variety of assessment tools and strategies are to be used to “gather relevant functional, developmental, and academic information, including information provided by the parent” (20 U.S.C. 1462 § 614(b)(2)(A)) to determine and assist in designing the most appropriate educational program for these students with disabilities. These accountability demands are promoting a greater emphasis on assessment and implementation of strategies that are directly related to producing higher achievement and in monitoring annual progress of these students.
(Fuchs, & Fuchs, 2002; Reschly, 2002; Shinn, 1998). Likewise, NCLB 2002 states that children with low incidence disabilities will typically fall in the group that will be those “students assessed with alternative assessment based on alternative achievement standards (‘proficient’ or ‘advanced’ scores on such tests limited to 1% of all children tested)” (70 Federal Register / Vol. 70, No. 240 /74631, Dec 15, 2005).

Both pieces of legislation now hold schools accountable for the academic achievement of all students, including those with the most significant cognitive disabilities (68 Federal Register / Vol. 68, No. 236 /68698, Dec. 9, 2003). Students with low-incidence disabilities are to be included in an assessment that is related to the state’s established academic content standards. However, an accountability process that measures the progress of these children may include different assessment techniques than those used in the general population. Those with the most significant cognitive disabilities are “measured against alternative achievement standards aligned with the state’s academic content standards and reflecting the professional judgment of the highest learning standards possible for the student” (Congressional Research Service, 2005, p. 5-6). IDEA 2004 and NCLB requirements require states to develop an accountability system that addresses modified achievement standards from the general education curricula requiring improvement in alternative means of assessment for those students with disabilities for whom the regular assessments, even with accommodations, are inappropriate (Turnbull, Turnbull, & Wehmeyer, 2007).

A strong concern for educators working with this low incidence population (that fit the criteria requiring alternative assessments) is the ability to design an effective educational program based on each individual’s strengths and needs. Local Education Agencies (LEAs) are required to demonstrate progress for these students based on individual goals and objectives “modified in such a manner that they reflect reduced breadth or depth of grade level content” (70 Federal Register / Vol. 70, No. 240 /74626, Dec 15, 2005), rather than based on grade-level standards. IDEA 2004 requires goals and short-term objectives that include academic (from the general curricula), functional skills and challenging behaviors (including identification of positive behavioral supports). Assessments, therefore, must appropriately identify the present level of performance in these given areas to facilitate the development of intervention strategies; this is a considerable change from past practice of merely creating goals and benchmarks related to scores on evaluations. In essence, professionals must provide an Alternative Means to Assess Skill Level (AMASL).

Results of the assessment process should lead to a useful Individual Family Service Plan (IFSP) or Individualized Educational Program (IEP) that can be implemented efficiently and effectively in both the home and classroom. The IFSP/IEP should allow both parents and teachers to directly measure the effectiveness of their child’s instruction and intervention strategies by observing the child’s progress toward his/her overall outcomes. Appropriate goals and objectives, if developed based on the “actual” knowledge and “true” skills of the student, will better measure and monitor the student progress, as well as his/her overall outcomes’ performance. The AMASL process, if implemented in practice, should and would correctly identify the appropriate goals and objectives necessary in reaching overall performance outcomes, and should be directly related to producing higher achievement and in monitoring progress of these students. The assessment process then becomes a means by which to provide an alternative measure of assessing the child’s skill level and performance progress that is directly linked to the IEP or IFSP.

IDEA 2004 strongly emphasizes that the information provided by the parent is to be used to determine and assist in designing their child’s educational program. This emphasis on parental direct and active involvement has long been a “best practices” philosophy of providing early intervention to those families and youngsters served under both Part B and Part C programs. Parents are identified as an integral part of the evaluation and IFSP/IEP teams; information from the parents must be included as part of the evaluation, making them an integral part of their child’s educational planning. Including parents of those children with low incidence disabilities in the
assessment process is even more important because they usually are more familiar with their child’s particular modes of communication, patterns of learning, and social nuances which are specific to that child. Professionals, on the other hand, can only reach the parents’ level of knowledge about the child after working with the child intimately over a lengthy period of time. The use of parent input and participation can eliminate wasting valuable instructional time as a result of the professional’s lack of knowledge about the child.

Thus, a combined approach, using both professional expertise and parent knowledge of their child, would seem to be the most effective means to obtain the necessary and critical information in both formal and informal assessments. Integrating these combined information sources should provide a more accurate “present level of performance” picture of that child. In fact, using only one type of measure is neither legal nor sufficient to obtain the necessary kinds of information required to design, develop and then provide an appropriate education program. Specifically designed program planning based on accurate evaluation information is necessary to ensure that these children with low incidence disabilities receive the most effective IFSP or IEP, as well as effective educational programming using the most appropriate intervention strategies.

It is those combined “best practice strategies” that the authors are suggesting be used to develop a more accountable educational program through the implementation of effective interventions that are directly related to producing higher achievement of specific skills and in monitoring progress of the children with low incidences. In order to determine these children’s overall performance outcomes, one must have correctly determined the child’s “true” present level of performance. From this information a determination of appropriate and specific educational program goals can be made that are the “actual” skills the child has displayed throughout the evaluation and assessment process, and not on the child’s overall performance based merely on scores constrained by basal and ceiling levels. Through the design and development of specialized instruction, one can efficiently evaluate the child’s progress by monitoring specific benchmark performance of various skills that were gleaned from what the child “actually did” and NOT based on what the child “could not do” as determined by conventional scoring procedures (e.g., basal and ceilings).

Overview of Cri-du-chat Syndrome

Cri-du-chat Syndrome (CDCS) is a genetic syndrome caused by a deletion of a portion of the short arm of the fifth chromosome. With prevalence rate estimated at about 1 in every 35,000 live births CDCS (M. E. Carlin, personal communication, August 11, 2003) falls in the low incidence disability category. Children with CDCS would most often be included in the mental retardation category of low incidence disabilities. Overall, individuals with low incidence disabilities account for less than 1% of all children served under IDEA as reported in the 25th Annual Report to Congress on the Individuals with Disabilities Education Act (U. S. Department of Education, 2003). It is unknown what portion of that one percent would be individuals with CDCS.

Individuals with CDCS typically fall within the severe range of development. Their common physical features are as pronounced in individuals with CDCS as they are for other syndromes (i.e., Down Syndrome). CDCS distinguishable facial features include (a) an asymmetrical face described as moon shaped with a small jaw (micrognathia), (b) a small head circumference (microcephaly), (c) down slanting eyes (anti-mongoloid slant), (d) wide set eyes (hypertelorism), (e) and folds of skin in the inner eye corners (epicanthal folds) (Carlin, 1983, 1990; Niebuhr, 1978; Radmer, Bodurtha, Jackson-Cook, Brown, & Wilkins-Huang; 1993; Wilkins, Brown, & Wolf, 1980; Wilkins, Brown, Nance & Wolf, 1983).

Communication and language development of those individuals with CDCS is generally significantly delayed, resulting in these children having very little speech. Their unusual cat-like cry (which gives the syndrome its name) does not have any conclusive research that indicates the cry is a means of specific communication. The majority of these children have limited verbal speech, and many may use a modified sign language, pic-
ture symbols, and alternative communication modes (or any combination). It is not unusual for parents and family members to have to interpret the communication of individuals with CDCS for others. Receptive language abilities are more advanced than expressive language. Overall, most if not all individuals with CDCS have communication issues ranging from mild to very severe. Although not a diagnostic criteria, communication and language issues arguably are one of the most critical concerns in development (Campbell, Carlin, Justen, & Baird, 2004).

Research indicates that individuals with CDCS have a wide range of problem behaviors (Carlin, 1990; Collins & Cornish, 2001; Dykens & Clarke, 1997). Behavior concerns are common, often centering on attention problems (i.e. hyperactivity, short attention spans). There has been some research documenting concerns with aggressive behaviors in a few individuals with CDCS. Nonetheless, individuals with CDCS have been described as friendly and affectionate (e.g. Carlin, 1990). Individuals with CDCS also have some physical concerns. These include low muscle tone, hyper sensitivity to sounds, reflux, frequent ear infections, and difficulty with feeding.

The developmental concerns in cognition, communication, behavior, and health/physical have implications in performing a valid evaluation or assessment of these children. Although children with CDCS share many commonalities, they also exhibit a wide range of ability levels. The ability level of these children runs along a continuum of significant to mild, making accurate assessment of their skill level especially important.

**Purpose of the Study**

This study looked at the overall collection of evaluations and assessments information as reported by both parents and professionals in regards to the overall development of young children with a low incidence disability. We reviewed evaluation and assessment information gathered on a number of young children with low-incidence disabilities (specifically, those diagnosed with Cri-du-chat) across various states. This collection of evaluation information indicated a strong variance in the development and evaluation of assessments procedures including the effective evaluation of their overall skills, the accurate assessment of their “true” present level of performance, and the means of which to assess the appropriate modalities of the data collection for determining progress of these children with low incidence. The means used to determine progress of the particular individuals’ educational program plan, commonly were evaluation instruments with generic basal and ceiling levels without considering splinter skills which is common in these children. In essence, one is trying to measure the skill level of children using a norm referenced instrument, when the norming group for which the instrument was developed is not representative of these children.

The research on low incidence and significant disabilities indicates that these individuals demonstrate inconsistent and various skill levels that are scattered across each of the developmental domains. It is the authors’ experiences that typically, these individuals’ cognitive skills and testing scores do not reflect their actual overall abilities. Therefore, traditional intelligence testing indicates a very poor prognosis for these individuals. Yet the experience of the authors and this data indicates that these individuals demonstrate a much better functional level than determined by standardized assessments. Since eligibility must be documented, standardized instruments are important. Practitioners, however, need additional information to design effective educational programs for children with low incidence and significant disabilities. In this study the authors will specifically address those children with the low incidence disability of Cri-du-chat Syndrome. The incidence rate is probably about 1 in 35,000 (Campbell et al., 2004).

**Method**

**Procedure**

Data were gathered over a two-year period at the annual conferences of the Five P Minus Society, the parent organization for individuals with Cri-du-chat Syndrome (CDCS). The two meetings were in Memphis, TN in August 2003 and Philadelphia, PA in August 2004. University master level graduate students per-
formed the assessments under the supervision of the authors. In this way, reliability concerns of the assessment procedures were lessened. Assessments were performed at the hotels where CDCS conferences were held. Parents reported on their child’s development using the Developmental Observation Checklist System (DOCS), and they were interviewed to compare assessment findings.

This research sample is one of convenience and cannot be considered a representative sample of individuals with Cri-du-chat Syndrome. These families participating in the study are actively involved in gaining the best services for their children, as well as having the financial ability to attend these international family support conferences. Since children with CDCS are such a low incidence population, this strategy was the most effective means to gather data on many children with the syndrome. The sample is large compared to other studies of children with CDCS, and the abilities of these children are consistent with published research (e.g. Baird, Campbell, Ingram & Gomez, 2001; Carlin 1983, 1990).

The assessment team administered a Battelle Developmental Inventory (BDI; Newborg, Stock, Wnek, Guidubaldi, & Svinicki, 1984) a DOCS, (Hresco, Miguel, Sherbenou, & Burton, 1994), a Scales of Independent Behavior Revised (SIB-R, Bruininks, Woodcock, Weatherman, & Hill, 1984), a Temperament and Atypical Behavior Scale (TABS, Neisworth, Bagnato, Salvia, & Hunt, 1999), and a video taping of each child to compile a language sample. Parents reported on types, frequency, and effectiveness of educational supports for the previous three years. Parents also reported on integration with typical peers.

Participants were informed of the study through a mailing prior to the meeting, and then recruited by the authors at the meeting. Parents completed the DOCS, SIB-R, TAB, and demographic sheets on their children. Appointments were made for each family with an assessment team. The team then administered the BDI.

**Instruments**

The BDI is a standardized, norm-referenced test that summarizes the key developmental skills in children ages birth to eight years. The BDI produces standard scores that allow the child’s development to be compared to the scores of a child’s typically developing peers. The BDI tests five domains: adaptive, motor, communication, cognitive, and personal-social.

The Developmental Checklist section of the DOCS was completed to acquire information about each child’s performance in his/her natural environment. The DOCS is a standardized, norm-referenced instrument completed by the child’s parent or primary caregiver or anyone familiar with the child’s development. This instrument provides information concerning a child’s everyday behavior. The DOCS measures a child’s development between the ages birth through six years, and includes the social, motor, language, and cognition domains.

The TABS is a reliable and valid norm referenced individually administered checklist of dysfunctional behavior used for children aged 11 to 71 months. The instrument measures temperament that the authors define as the child’s characteristic emotional style or disposition. The TABS is used to identify atypical behaviors that are found to possibly threaten present or future development.

The SIB-R is a comprehensive norm referenced measure of adaptive and problem behaviors. It is designed for early infancy to 80 years of age. A particular strength of the instrument is the problem behavior section, which assesses these behaviors along with their frequency and intensity.

**Participants**

Twenty seven individuals with the low incidence disability of CDCS from three to 18 years of age and their families participated in Memphis, TN (8/2003) study. Of the 27 participants, 18 were girls and nine were boys. Eight individuals with CDCS and their families participated in Philadelphia, PA (8/2004) study, five girls and three boys. A total of 35 individuals with CDCS were included.

**Data Analysis**

The statistical analysis between the parent report from the DOCS and the BDI were mea-
measured with correlations of the paired domain variables. For example the DOCS Social domain was correlated with the BDI Personal Social. We then used a paired sample t-test again comparing the domains from parent report to administered assessment results. Finally, we used multiple regression to test if the four domains and overall score on the DOCS (Independent Variables) would be a predictor of the overall development score on the BDI (dependent variable). In this way we were able to make comparisons both across domains and with overall development. The t-test further specified if the means were significantly different.

Results

Using Cook’s Distance analysis we eliminated one case that was shown to be an outlier. The parent report for this child was very different from the BDI results. Removing this outlier strengthened the correlation results, but only by a factor of less than .03 across the five pairs. Table 1 reports descriptive statistics of the BDI and DOCS results with an adjusted n of 34 from eliminating this case. The table displays the range of abilities for these children across domains. The table also includes the minimum and maximum along with the mean and standard deviation. The minimum and maximum are the scores, reported in age equivalents, by the respective instruments. The wide range in these scores is not surprising given the wide range in abilities in this population. Mean scores for this group are indicative of the high level of delay given the mean age for this group of almost 108 months. Comparisons of scores across age groups are described in Table 2. There were statically significant

<table>
<thead>
<tr>
<th>Domain</th>
<th>Instrument</th>
<th>Minimum</th>
<th>Maximum</th>
<th>Mean</th>
<th>SD</th>
</tr>
</thead>
<tbody>
<tr>
<td>Personal Social</td>
<td>BDI</td>
<td>1</td>
<td>74</td>
<td>36.51</td>
<td>16.64</td>
</tr>
<tr>
<td></td>
<td>DOCS</td>
<td>11</td>
<td>72</td>
<td>42.40</td>
<td>18.43</td>
</tr>
<tr>
<td>Motor</td>
<td>BDI</td>
<td>8</td>
<td>83</td>
<td>32.51</td>
<td>17.05</td>
</tr>
<tr>
<td></td>
<td>DOCS</td>
<td>11</td>
<td>72</td>
<td>40.86</td>
<td>17.49</td>
</tr>
<tr>
<td>Communication</td>
<td>BDI</td>
<td>0</td>
<td>74</td>
<td>28.34</td>
<td>16.72</td>
</tr>
<tr>
<td></td>
<td>DOCS</td>
<td>10</td>
<td>72</td>
<td>41.66</td>
<td>18.87</td>
</tr>
<tr>
<td>Overall</td>
<td>BDI</td>
<td>4</td>
<td>69</td>
<td>34.49</td>
<td>15.01</td>
</tr>
<tr>
<td></td>
<td>DOCS</td>
<td>11</td>
<td>72</td>
<td>40.97</td>
<td>17.78</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Table 2</th>
<th>Post Hoc Results: Mean Variances Comparison by Age Group, Descriptive Statistics for Participants (n = 34) (Significant findings)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Instrument Domain</td>
<td>Age range 1 (months)</td>
</tr>
<tr>
<td>BDI Personal Social</td>
<td>73 to 144</td>
</tr>
<tr>
<td>BDI Cognition</td>
<td>0 to 72</td>
</tr>
<tr>
<td>BDI</td>
<td>0 to 72</td>
</tr>
<tr>
<td>Overall</td>
<td>73 to 144</td>
</tr>
<tr>
<td>DOCS Social</td>
<td>0 to 72</td>
</tr>
<tr>
<td>DOCS Motor</td>
<td>0 to 72</td>
</tr>
<tr>
<td>DOCS Language</td>
<td>0 to 72</td>
</tr>
<tr>
<td>DOCS Cognition</td>
<td>0 to 72</td>
</tr>
<tr>
<td>DOCS Overall</td>
<td>0 to 72</td>
</tr>
</tbody>
</table>
findings in several areas. The only statistically significant difference in gender was in the area of cognition which had a between group difference ($F_{11005} = 5.59$, $p = .024$) with a mean for boys of 46.58 months ($SD = 16.28$) and girls of 32.45 months ($SD = 16.88$). The reader should be cautious in interpretation due to the small sample size ($n = 34$) and especially for boys ($n = 12$) may contribute to this difference.

Using paired sample correlations, we compared results from the parent reported DOCS and the field administered the BDI. For this group of children, the parent’s reports of their child’s development were very congruent with the standardized results. As we expected the parent’s reported levels that are somewhat higher than those we report from the BDI results. The disparity is most likely because the examiners were unfamiliar to these children. Parents, as part of the process, did report that the assessment results were for the most part typical of their children’s abilities. Table 3 shows the paired sample correlations for the two groups by domains. The correlations show the relationship between results reported by the parents on the DOCS and assessment results in the BDI. We were able to compare corresponding domains from each instrument using overall and domain scores for the BDI in the areas where the DOCS does not discriminate sub-domains.

Although all of the relationships proved very strong, the researchers were particularly pleased with the very high score in Communication from the BDI versus Language from the DOCS. These children use a variety of communication modalities, including some words, sign language, augmentative communication devices, and gestures. The ability to accurately assess communication for these children with very little prior knowledge of the way they express themselves was encouraging. Parents of these children with low incidence disabilities are able to realistically gauge their children’s development even as they get older.

A t-test was used to assess the difference in the mean scores for the two groups. We did not find statistically significant differences in the BDI cognition / DOCS cognition. For the social domain, we did find a statistically significant difference ($t = 2.11$, $p = .042$) for motor ($t = 3.41$, $p = .002$), for BDI Communications / DOCS language ($t = 7.71$, $p < .001$), and for overall development ($t = 4.30$, $p = .001$). Given the lack of familiarity with these children and the short time frame to do assessment, this is not surprising. We were pleased to note there was no difference in cognition.

Finally, we used a multiple regression to see if the combined domains and overall score from the DOCS could predict overall development for these children. We combined the five variables from the DOCS, social, motor, language, cognition, and overall development to test if they were significantly related to the BDI. For the model we found an Adjusted R Square of .79, $p < .001$. We then removed the overall development variable from the equation to see if the composite score it represented might be inflating the statistic significance. With the DOCS overall score removed, we found an Adjusted R Square of .75 $p < .001$. The removal of the overall score from the DOCS in essence made virtually no difference in the prediction of BDI overall develop from the parents report from the DOCS.

**Discussion**

As indicated by IDEA 2004, a variety of assessment tools and strategies were used to gather relevant functional, developmental, and academic information, which included information provided by the parent of these children with the low incidence disability of Cri-du-chat Syndrome. These families are faced with some unique challenges, such as the significant variability of the overall development of these

<table>
<thead>
<tr>
<th>Domain</th>
<th>Correlation Coefficient</th>
<th>Significance</th>
</tr>
</thead>
<tbody>
<tr>
<td>Personal Social</td>
<td>.674</td>
<td>&lt;.001</td>
</tr>
<tr>
<td>Motor</td>
<td>.686</td>
<td>&lt;.001</td>
</tr>
<tr>
<td>Communication / Language</td>
<td>.861</td>
<td>&lt;.001</td>
</tr>
<tr>
<td>Cognition</td>
<td>.824</td>
<td>&lt;.001</td>
</tr>
<tr>
<td>Overall Development</td>
<td>.864</td>
<td>&lt;.001</td>
</tr>
</tbody>
</table>
children along with a very poor medical and educational professional prognosis.

These assessment results promoted a greater emphasis on assessment that can be directly related to producing higher achievement. The high correlation of parent and assessment results demonstrates that, for at least these children with low incidence disabilities, parents are able to provide professionals with an accurate portrayal of their child’s development. From this, better programs can be developed. Most importantly, parents can be equal partners in their child’s education and can assist in monitoring progress.

Although it has been documented previously that a strong relationship exists between parental report and assessment results (i.e., Dinnebeil & Rule, 1994), this study extends those findings to a significantly involved low incidence population. As accountability continues to be an important consideration in Early Childhood Special Education these findings offer IFSP/IEP teams the knowledge that team members have a similar understanding of the child’s ability that has been documented through multiple sources and instruments. Teams can feel more comfortable, as part of the evaluation assessment process, in accepting parent report, thus eliminating the need for at least some standardized assessments. This will allow programs to spend more time and resources on interventions that are more likely to be successful, rather than on additional evaluations.

For parents of children with Cri du chat Syndrome this information will assist them in working with professionals who are probably not familiar with the syndrome as they are seeking ways to help their children. Because this is such a low incidence disability, information is rare; recent medical and educational research indicates a much better prognosis that many professionals may not realize. The improved prognosis is due in part to more sophisticated genetic testing, which has identified many children with smaller deletions as having CDCS. Thus, children with smaller deletions are now being identified. The tendency in the syndrome is for smaller deletions to be less delayed; as a result, children with the smaller deletion are performing at much higher levels than is often expected by professionals. In addition, as a field we are better able to assist young children with significant disabilities to reach a greater potential than a few years ago. For all of these reasons, families are often challenged to get professionals to see the more advanced potential in their children.

With IDEA 2004 a new focus of accountability has emerged. Teachers and schools must show annual progress of their students. A realistic profile of the child is a key and critical piece needed to document change occurring through intervention. Time spent providing interventions that are not in tune with the child’s skill level may result in their not meeting that annual progress goal, either because of time lost in changing interventions or working on skills that are either too low or high for this child. Since children with CDCS have such poor communication it could be difficult to determine why the intervention may not be effective. For this reason having an accurate measure of ability at the outset of the intervention gives teachers and parents more confidence in their intervention strategies.

References


Received: 21 September 2006
Initial Acceptance: 5 December 2006
Final Acceptance: 20 July 2007