

Outcomes in Children who are Deaf-Blind with Cochlear Implants

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www.kidsdbci.org/

Background/Objective

- Rationale: Deaf-blind children are receiving cochlear implants, however little is known about outcomes in this complex group of children
- Objective: To describe language skills in children with dual sensory impairments with implants

Relevance

- Children with dual sensory impairment are receiving implants at increasing rates
- Little is known about how children are doing
- Often CI teams and EI providers have little training in how to effectively intervene for a deaf-blind child with an implant

Additional Disabilities in Children with SNHL

No additional disabilities	60.7 %
MR	8.7 %
Developmental Delay	4.8 %
Learning disability	8.3 %
ADHD	5.6 %
Blindness and Low Vision	5.2 %
Motor/CP	4.4 %
Emotional	2.0 %
Autism	1.6 %
Other	0.3 %

From 2007-2008 Gallaudet survey

Visual Impairments in Deaf/HOH

- Deaf children are 2-3 times more likely to develop vision problems than hearing peers (Guy et al, 2003)
 - 15.3% incidence of refractive errors hearing children
 - 39.1% in group of deaf children
- Rate of ophthalmologic findings 21.7% (Sharma et al 2009)
- Usher Syndrome (3 types)
- Should have a full ophthalmologic evaluation
- Need **regular** vision evaluations

Terminology of Vision Impairments

Legal blindness

- Central visual acuity of 20/200 or less in the better eye after correction or central visual acuity of more than 20 / 200 if there is a visual field defect in which the peripheral field is reduced to an angle of 20 degrees or less in the better eye (Koestler, 1976, p. 45)
- Partially sighted (or partially seeing): Visual acuity between 20/70 and 20/200 after correction in the better eye (Although this term is used in many state definitions, it is considered outdated and is not being used by practitioners)

Terminology of Vision Impairments

Visual acuity

- This refers to the ability to clearly distinguish forms, objects or symbols at specific distances (Gothelf, Rikhye, & Silberman, 1988)
- The term 20/200 means that the person with the visual impairment can see an object or symbol at 20 feet that a person with unimpaired vision can see at 200 feet

Field of vision

- The ability to see objects in the periphery of ones vision when looking straight ahead
- Individuals with unimpaired vision can usually see objects within 180 degree arc when looking straight ahead

Definitions used in Education

Visual Impairment (visual handicapped)

- This term encompasses a wide range of vision loss which can include deficits in acuity, field loss, ocular motility, or color perception which may be permanent or temporary
- The term visual handicapped is often used synonymously with the term visual impairment to refer to a vision loss which even with correction, adversely affects a child's educational performance (P.L. 94-142)

Definitions used in Education

Blind

- Individuals who are totally without vision or who have light perception only
- In the educational field, this term refers to children who use other senses (i.e. hearing and touch) as primary channels for learning or receiving information

Light perception only

- Individuals who are without vision, but can perceive light

Definitions used in Education

Low vision

- This is a broad term which is used to refer to individuals who have significant visual impairments, but still have usable vision
- Vision is still used as a primary channel for learning or receiving information
- Although the visual impairment continues after correction, visual functioning may increase with the use of optical aids, environmental modifications and/or training (Corn, 1980; Scholl, 1986)

Cortical Visual Impairment (Cortical Blindness)

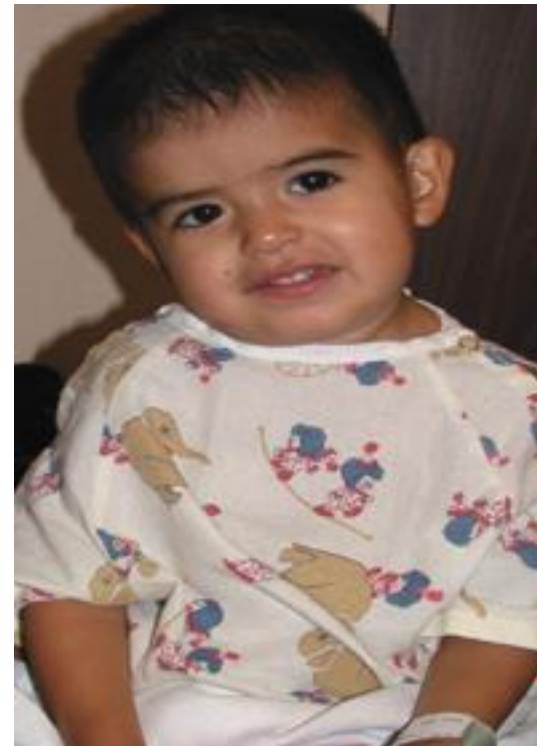
- The visual cortex of the brain or the visual pathways to the brain is damaged
- This results in varying visual impairments, depending upon the location of damage in the brain
- The anatomy and physiology of the eye is not impaired

Visual functioning

- This refers to how well a person uses the vision he or she has
- Visual functioning is considered a learned behavior which is not necessarily reflected by visual acuity. It is possible for a student to have poor visual acuity and good visual functioning, or visa versa (Gothelf, Rikhye & Silberman, 1988)
- Individuals who fail to use their vision in an efficient or meaningful way have poor visual functioning and are often included in functional definitions describing visual impairments

Red Flags for Vision Concerns

- Poor visual regard
- Poor tracking (up or down)
- Wiggling eyes
- Wandering eyes
- Head tilt



Risk factors for Hearing and Vision Problems

- Family history of vision problems
- Prematurity/NICU related problems
- Birth asphyxia
- Infections (congenital CMV, toxoplasmosis, rubella, meningitis)
- Traumatic brain injury
- Certain syndromes

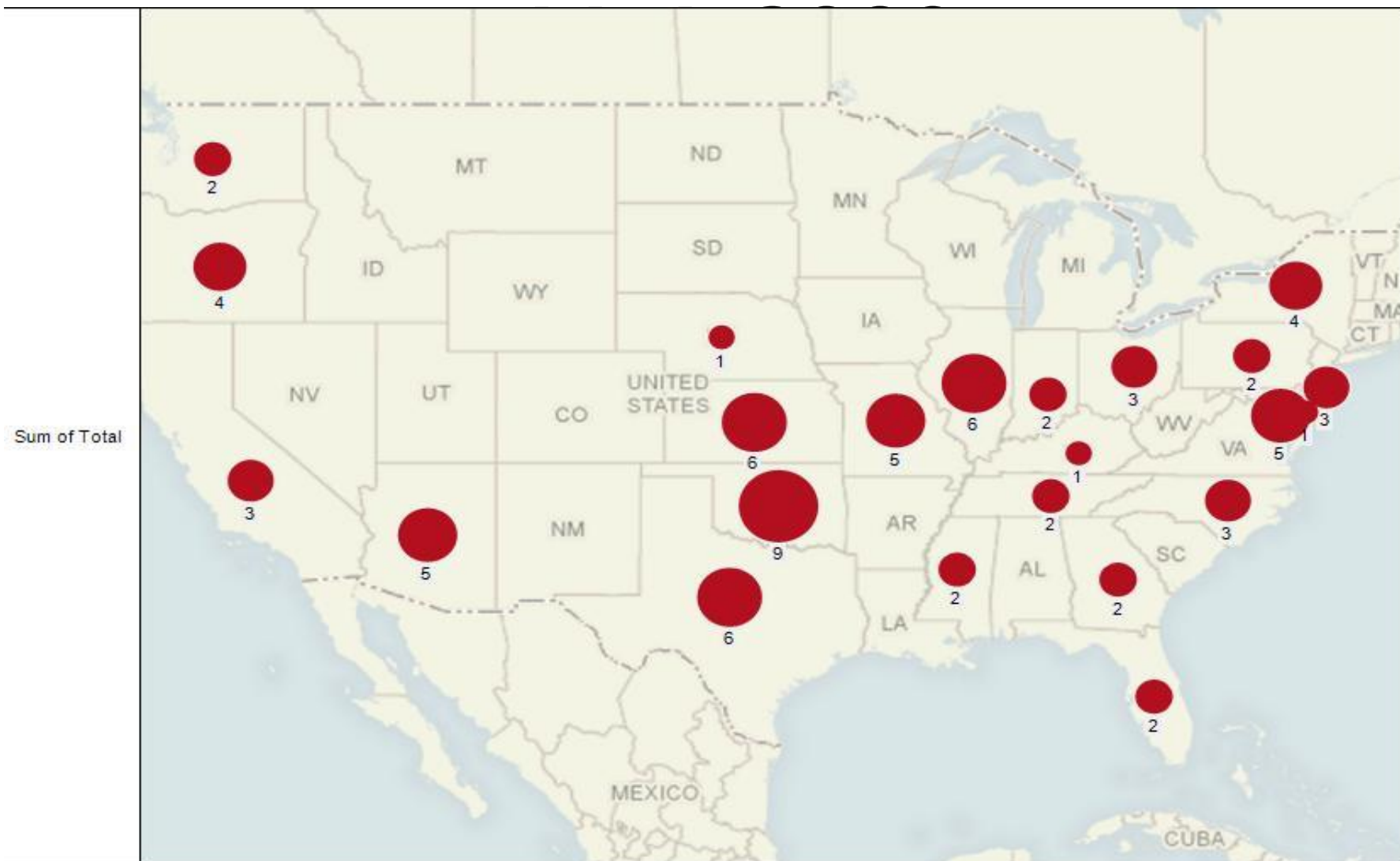
Syndromes associated with HL and vision impairment

- CHARGE Syndrome (CHARGE Syndrome Foundation www.chargesyndrome.org/)
- Usher Syndrome (www.usher-syndrome.org/)
- Stickler Syndrome
- Treacher Collins Syndrome
- Goldenhar Syndrome
- Infantile Refsum's

Study

- Multi-center study
- Enrolled deaf-blind children, 12 months to <6 years who had or will receive a cochlear implant
- Evaluated language outcomes
- Assessed language, development, auditory skills
 - Communication and Symbolic Behavior Scales
 - MacArthur-Bates Communication Scale
 - **Reynell-Zinkin (developmental assessment for DB)**
 - Infant-Toddler Meaningful Auditory Integration Scale or Meaningful Auditory Integration Scale (ITMAIS-MAIS)
 - Speech Intelligibility Rating

Locations of Study Subjects



■ Texas Deafblind Project *

■ Project Staff or Consultants conduct assessments, not State Project staff

Reynell Zinkin: Response to Sound

Detection Skills	<p>1: Listens to sounds</p> <p>2: Orientation towards sound of source</p> <p>3: Selective response to sound</p> <p>4: Reaching for source of sound in any direction</p> <p>5: Reaching for source of sound in correct direction</p> <p>6: Recognition of familiar sounds (own toys, parents voices)</p>
Response to simple words/phrases	<p>7: Recognition of familiar words or phrases</p> <p>8: Appropriate response to familiar phrase or words</p> <p>9: Appropriate response to simple direction (give it to me, give it to mommy, where is your nose)</p> <p>10: Selection of familiar object in response to naming</p> <p>11-15: selection of objects from choice of 3</p>
Word Identification	<p>10: Selection of familiar object in response to naming</p> <p>11-15: selection of objects from choice of 3</p>
Simple Directions	<p>16-20: directional commands with items (put the spoon in the cup)</p>
Understands functional use of objects, spatial concepts, size	<p>21-36:</p> <p>Which one do we drink out of, Find two things we can use for eating dinner, Show me the longest pencil, the smallest cup</p> <p>Put the short pencil in the biggest cup</p>

Vocalization and Expressive Language

Sound Production	Some meaningful words
1: vocalization other than crying	9: 2-3 meaningful words
2: single-syllable sound	10: 4-6 words
3: two different sounds	11: 6-12 words
4: four different sounds, including consonants	12: word combinations
5: double syllable sounds	13: 20 or more words
6: repetitive double syllable babble	Sentences
Word	14: appropriate use of words other than nouns or verbs
7: one definite words	15: Sentences of 3 or more words
Expressive jargon	16: Appropriate use of prepositions
8: expressive jargon	17: appropriate use of pronouns

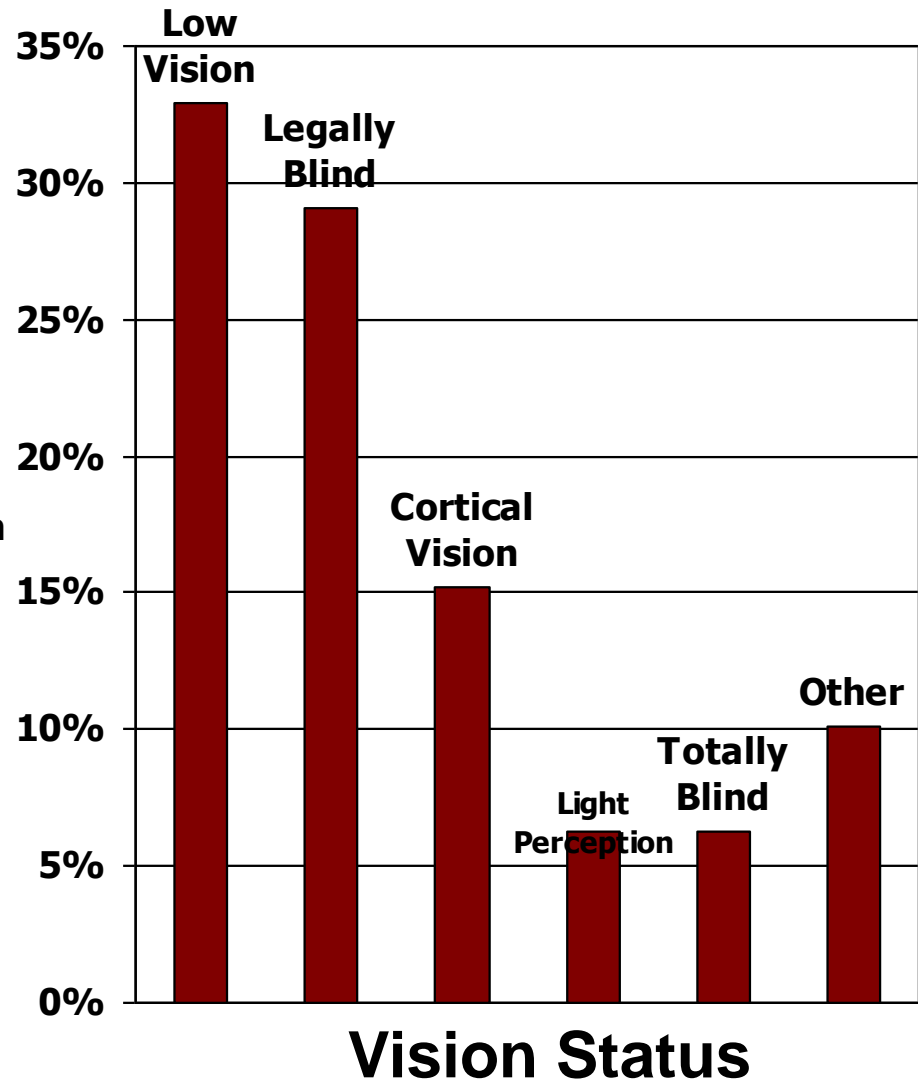
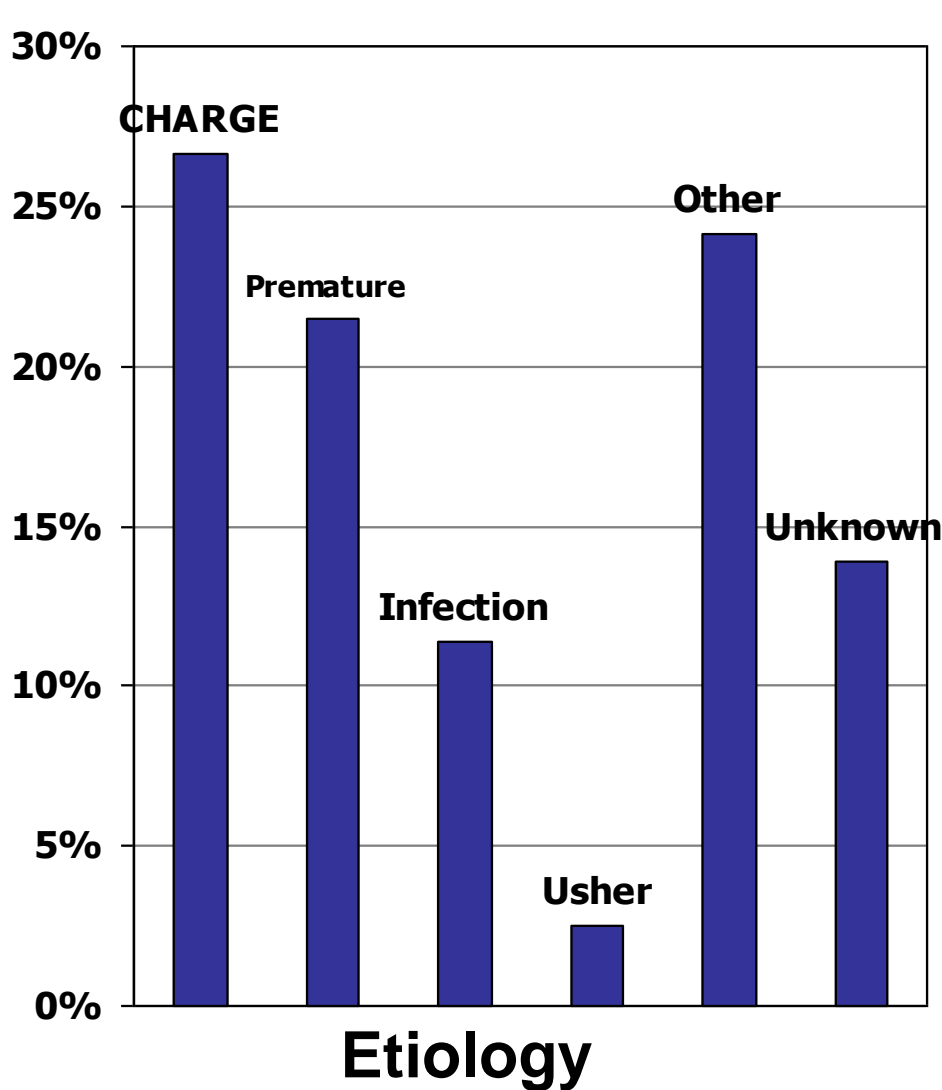
Vocalization and Expressive Language

Complex sentences
18: any appropriate use of past tense
19: mature forms of past tense
20: appropriate use of future tense
21: nearly all sentences correct and complete
22: use of complex sentences

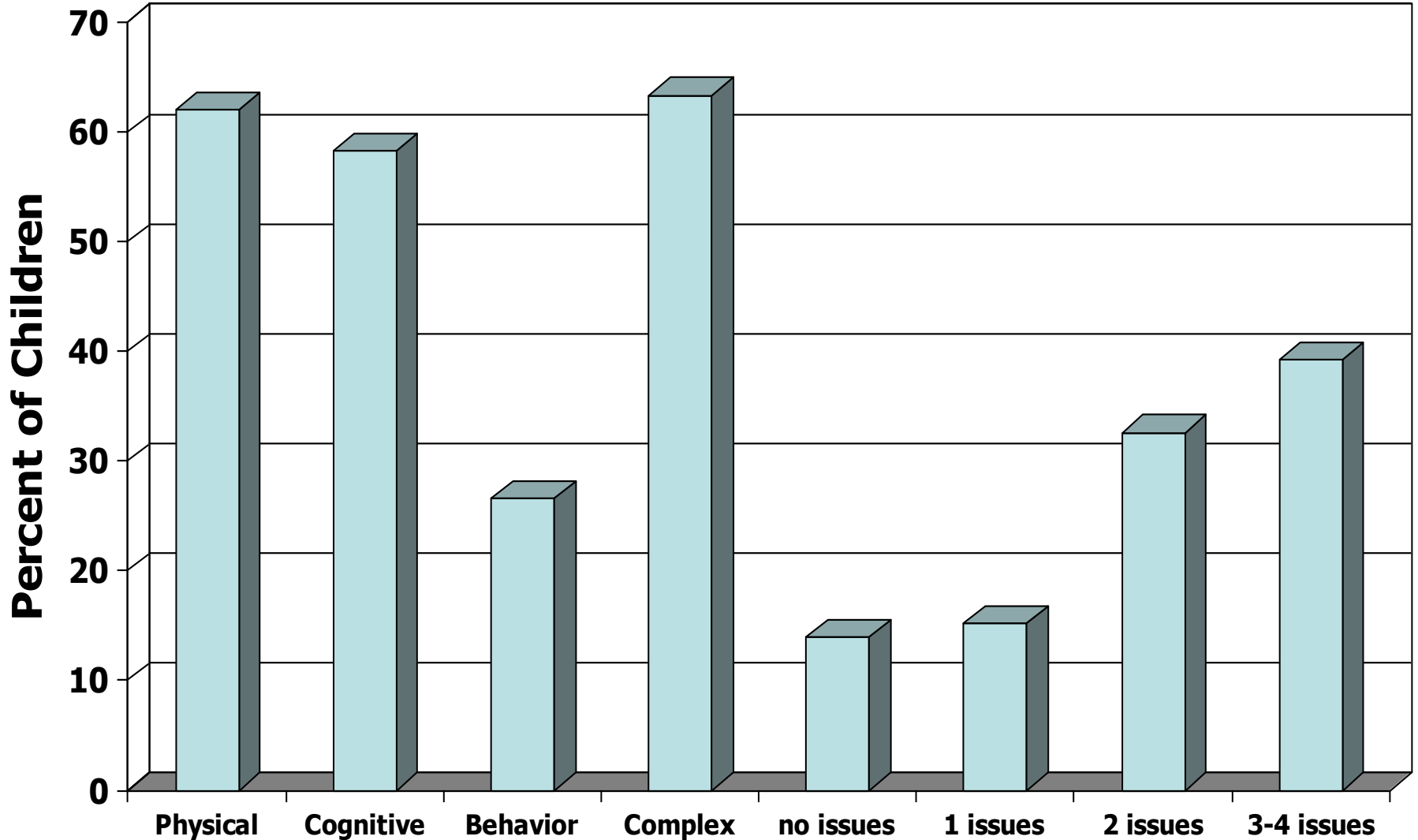
Subject Characteristics

Characteristic	N=79
Median age in months at study enrollment	48 (12-96)
Median age in months at last study visit	64 (19-107)
Median age in months at 1st implant	22 (6-74)
Median duration (in mos) with 1st implant at last study visit	36 (3-83)
Bilateral implants	21% (17)
Ethnicity	
Caucasian	75% (59)
African American	6% (5)
Latino	11% (9)
Other	8% (6)
Median developmental ability in months at last study visit	15 (1-36)

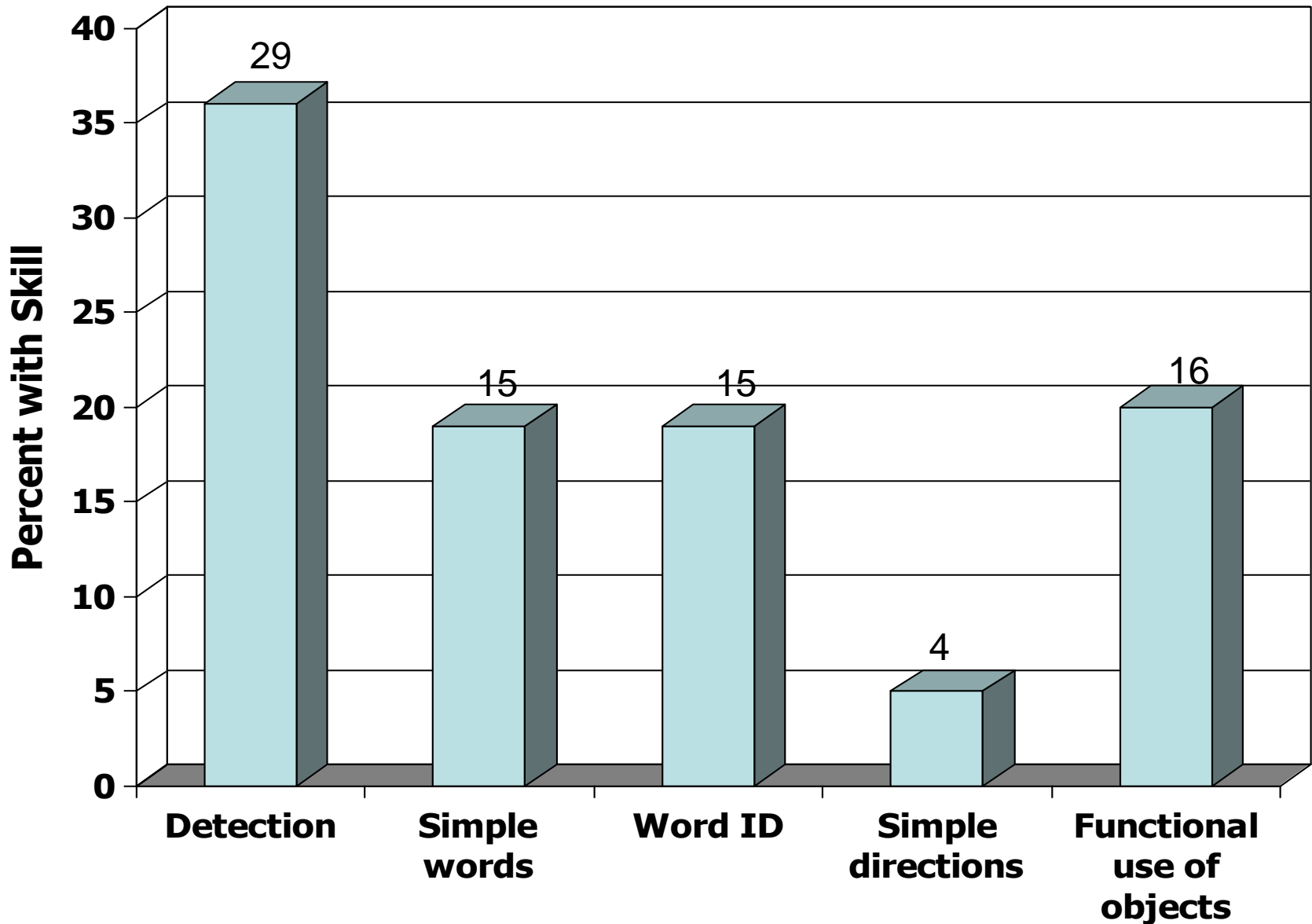
Characteristics



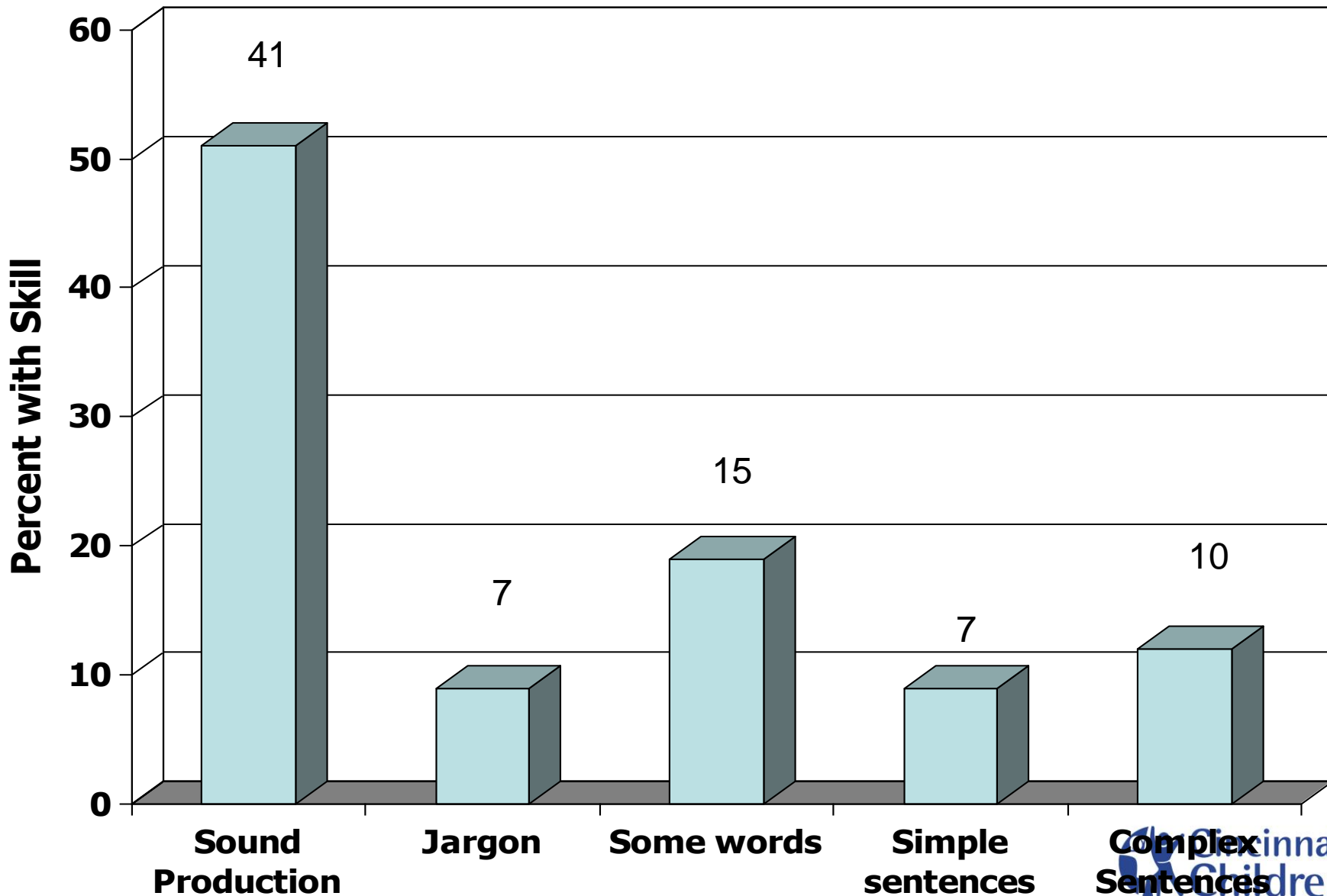
Additional Developmental Issues



Reynell Zinkin Response to Sound



Reynell Zinkin Expressive Language



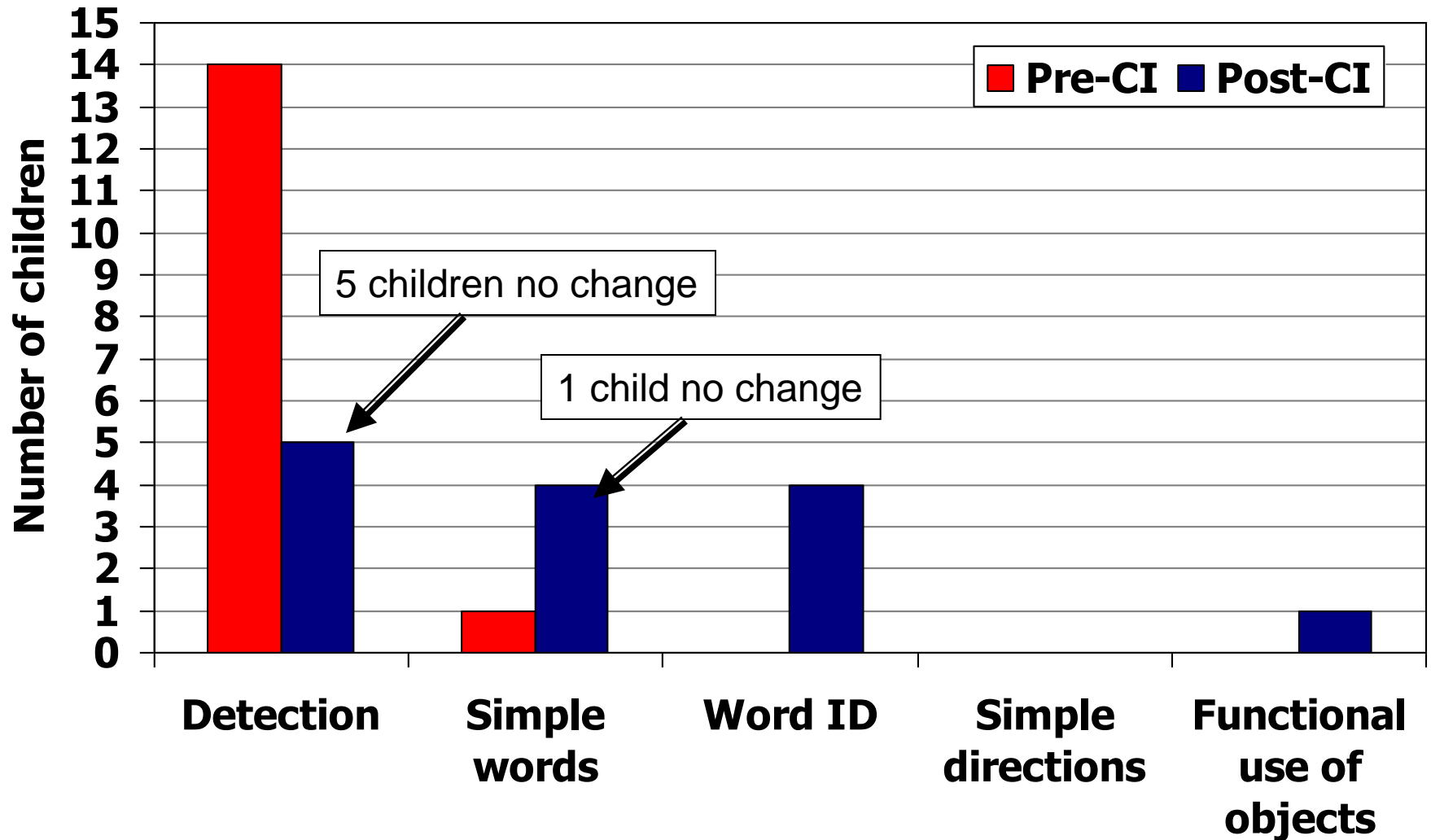
Reynell Zinkin

- **Age at implant** was **not** correlated with receptive ($\rho=-.020$, $p=.86$) or expressive ($\rho=.147$, $p=.19$) language outcomes
- **Duration with implant** was significant, but not strongly correlated with receptive language ($\rho=.310$, $p=.005$) or expressive language ($\rho=.372$, $p=.0001$) outcomes
- **Cognitive level** showed good correlation and significance with receptive language ($\rho=0.66$, $p<.0001$) and expressive language ($\rho=.619$, $p=.001$)

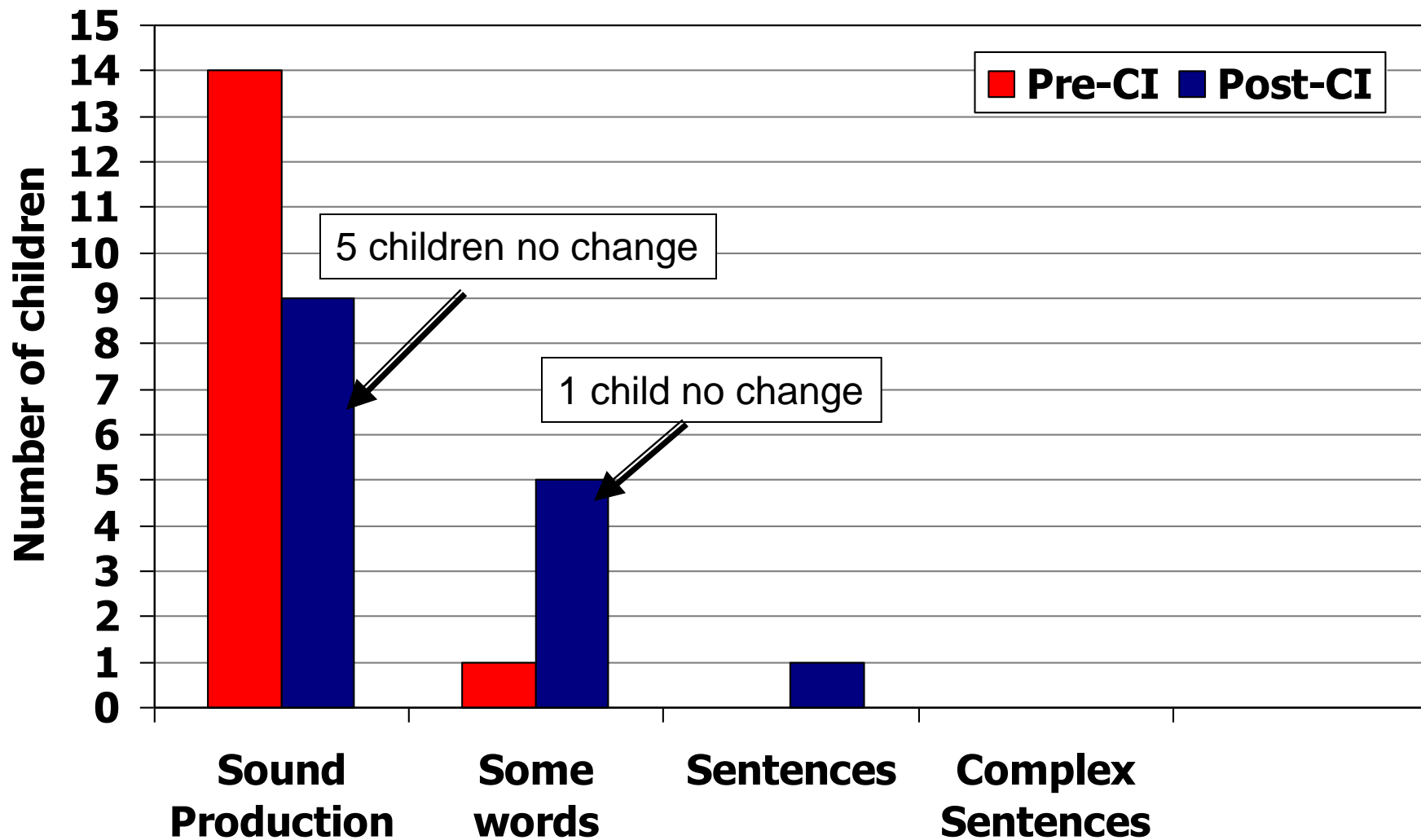
Subject Characteristics

Characteristic	N=15
Median age in months at pre-CI visit	20 (12-70)
Median age in months at 1 st implant	22 (12-72)
Median duration (in mos) with 1 st implant at last study visit	12 (3-37)
Vision Status	
Low Vision	47% (7)
Legally Blind	13% (2)
Light Perception	6% (1)
Totally Blind	27% (4)
Other	6% (1)
Disability Type	
Physical	60% (9)
Cognitive Impairment	54% (8)
Behavior Problem	13% (3)
Complex Medical condition	54% (8)

Change in receptive language over time



Change in expressive language over time



How do the usual predictors fare?

- **Age at implant** was not correlated with post-implant
 - receptive language level ($\rho = 0.22$, $p = .939$)
 - expressive language level ($\rho = -.147$, $p = .60$)
- **Duration of implant** showed a stronger but not significant correlation with
 - Receptive language level ($\rho = .567$, $p = .028$)
- **Duration of implant** was not correlated with
 - Expressive language level ($\rho = -.128$, $p = .65$)

How do the usual predictors fare?

- **Developmental quotient** was correlated but not to a statistically significant level
 - Post-implant receptive language skills ($\rho = .458$, $p=0.86$)
- **Developmental quotient** was correlated and reached statistical significance for
 - Post-implant expressive language skills ($\rho = 0.570$, $p=0.026$)

Correlates of Language

- Cognitive level significantly correlated with receptive language ($\rho=0.66, p<.0001$)

Intervention

- Functional Vision Assessment
- Adaptations
- Routine-based interventions
- Resources

Functional Vision Assessment

- A teacher of the visually impaired is essential in the provision of services to children with dual sensory impairment
- The eye exam/ophthalmology exam only gives limited information about vision
- Getting a sense of how a child uses their vision and the best approach to provide information is critical

Functional Vision Assessment

- May assist you in determining:
 - Best lighting (light focused on the item, backlighting with a light box, etc)
 - Best angle or presentation of information
 - Best font size/contrast needs
 - Best speed with which we can present information (visual tracking)
 - Most visually relevant information for the child
 - Tactile adaptation of materials

General Adaptations for Children Who are Deafblind (Pam Shanks)

- May need extra time to process information, young children may need up to 15 seconds (Miles, 1999)
- Present information within a range that allows the child to perceive it
- Use touch cues and object cues that are distinctive prompts made on a child's body to convey information and anticipation (eg., this is mommy, bottle on tummy)

General Adaptations for Children Who are Deafblind (Pam Shanks)

- Present information consistently
- Present information slowly
- Wait for reactions that indicate perception or understanding
- Look for anticipation in the child that may be subtle but suggests understanding

Routine-Based Intervention

- Determine sounds that may be relevant in the environment
- Identify routines that are salient to family and child
- Work towards increasing hierarchy of communication skills

Sound Inventory (Stremel, Workman)

Directions: Please check the sounds that are in your environment (on the left-hand side). Columns on the right-hand side can be used as an assessment to determine your child's detection and identification of specific sounds.

Sounds in Your Home & Community Environment	Your Child's Response to the Sounds		Motivating Sounds to Target for Learning
	Detection	Identification	
Home Environment – Kitchen and Utility Room:			
<input type="checkbox"/> Microwave bell	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
<input type="checkbox"/> Oven door opening/closing	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
<input type="checkbox"/> Oven/egg timer	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
<input type="checkbox"/> Oven temperature setting (beeps)	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
<input type="checkbox"/> Refrigerator opening/closing	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
<input type="checkbox"/> Drawer opening/closing	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
<input type="checkbox"/> Dishwasher	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
<input type="checkbox"/> Toast popping up in toaster	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
<input type="checkbox"/> Blender/Food Processor	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
<input type="checkbox"/> Electric can opener	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
<input type="checkbox"/> Garbage disposal	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
<input type="checkbox"/> Washing machine	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>

Hierarchy of Communication

Analysis of CI Assessments: Levels of Communication Ma

LEVEL	Behaviors
Prelinguistic	Makes wants known by any communication
	Responds to simple gestures
	Uses deictic gestures
	Uses representational gestures (Iconic signs)
Expressive Communication	Uses sounds to communication pleasure/dis
	Imitates vowel sounds/words
	Demonstrates ability to use a few syllables CV/VC
	Demonstrates ability to use a few words (3-5)
	Uses at least 50 words
	Joins 2-3 words together
	Uses modifying words
	Uses morphemes
Uses complex sentences	

Hierarchy of Communication

Comprehension	Detects sounds
	Responds to name
	Responds to Simple Phrases
	Demonstrates Object ID
	Demonstrates directives with verb-object-location/object
	Complex directions
Functions of Communication	Protest/rejection
	Request object/action
	Greet/Gets attention
	Shows objects/comments

Gestural Development Assessment

STEPPINGSTONES KIDSDBCI
 Gestural Development Assessment
 Stremel Thomas, 2010

⊕ Assessments Dates

Months					
Behavior regulation 9-12 Months	<u>Protest:</u> Child uses body to signal refusal/protest (e.g., arching body away when held in adult's arms Child pushes away an object with hand(s) <u>Requests objects:</u> Child points to obtain an object Child reaches for an object Child makes contact with an adult's hand to gain object <u>Request actions:</u> Child reaches to be picked up Child does an action to get it to happen again (e.g., bounces up and down for "horsie")				
Social	<u>Seek attention:</u>				

Gestural Development Assessment

Joint attention 15-18 months	<p><u>Comment:</u> Child points to an object in response to an adult's request, such as "Show me the apple" or "Where's the doggie?"</p> <p><u>Request information:</u> Child points to object or event to gain information (e.g., child points to pictures in book for adult to name it)</p>				
Social interaction 18-24 months	<p><u>Seeks attention:</u> Child shows off (e.g., sticks out tongue, makes a funny face to get a laugh)</p> <p><u>Representational gestures:</u> Child shrugs shoulders or puts hands face-up for "All gone" or "Where did it go?" Child blows kisses to others Child signals "shh" with fingers to lips Child nods "yes" Child pretends to sleep with hands together by head Child uses conventional gesture of excitement (e.g., "high five" or "touchdown")</p>				
Joint attention 18-24 months	<p><u>Comment:</u> Child uses gesture as clarification of word/word approximation (e.g., child says "pane" and then points to airplane when not understood)</p>				

Stremel Thomas, K. (2010) Adapted from: ~~Crais, Watson & Barnnek~~ (2009) American Journal of Speech-Language Pathology, Vol 1895-108. □

Scoring: 0 = Not Yet; 1 = Inconsistently; 2 = Consistently Uses

Routine-Based Worksheet

Developing Routines: A Worksheet

Name:

Date:

Routine: Bath time Major Purpose: to provide non-speech cues to provide information and opportunities for communication and partial participation.

Major Intervention Strategies: Pair touch and object with cues with verbal

Vocabulary Used in Routine: Toothbrush, bath, water, washcloth, wash, body parts, off, in, out, lotions, socks, diaper, arm, leg, tummy, face

Routines Based Worksheet

What you do!	R	E	M	DL	Cog	Support & AT	Targeted Outcomes for the Child	Consequences/ Expansions
	Comm.							
Getting Up & tooth brushing precede								
BEGINNING						Simultaneously give touch /object cue		
Run bath water								
Say C's name while giving touch cue on his hand	✓					Touch cue	*C responds by smiling or changing behavior	Provide verbal & touch consequence
Announce who you are/touch cue	✓					Touch cue		
Describe beginning step in the undressing routine	✓					Jiggle the snap/zipper	C could reach up to his snap	Tap him if he does this
Announce each step "arm out"	✓		✓	✓		Jiggle the arm cloth	C cooperates in undressing	Kiss his hand when arms are out
" " "leg out"	✓					Jiggle the leg cloth	C cooperates in undressing	Rub his legs when legs are out
" " "Socks off"	✓		✓	✓		Touch foot	C cooperates in undressing	
Announce diaper off	✓					Touch cue for diaper off		Show diaper and gesture, "Throw away"
Say, "Time for Bath"	✓		✓			Give wet washcloth for "bath" or a bowl to feel "water"	*C grasps and holds washcloth	Clap and smile in his line of vision

DB Resources

NATIONAL CONSORTIUM ON DEAF-BLINDNESS

Home
About NCDB
Contact NCDB

News and Announcements
About Deaf-Blindness
Text Only Version
en Español

For Families

Technical Assistance

Personnel Development

DB-LINK Info Services

- Selected Topics
- DB-Library
- People and Programs
- NCDB Products
- Conferences and Trainings
- Research to Practice
- Deaf-Blind Perspectives
- New Publications

–<http://nationaldb.org/>

National Consortium on Deaf-Blindness

- DB-LINK: many good articles, references
- Deaf-Blind Perspectives: a newsletter
- Up to date list of conferences and professional development workshops
- Family page: stories, articles
 - I wish I had known about non-verbal ways to communicate with my daughter Sara when she was much younger. When Sara was 10, we were introduced to a wonderful educator who specialized in non-verbal communication. Dr. Mary Morse came to Sara's class to talk to the school team about object communication and calendar boxes. It changed our lives for the better. Sara learned that objects represented activities and she loved the power that this knowledge gave her in school and at home.
 - ~ Janette Peracchio, Connecticut

<http://www.nationaldb.org/FFWish.php> (6 minute video)

Perkins School for the Blind



BY COLOR

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Vision Loss

Teaching
Resources

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Teaching Resources

Curricular Resources

Educational Publications

DB Guidelines

Resource Packets

Samuel P. Hayes Research Library

Scout: Information on Blindness &
Visual Impairment

Deafblindness: Educational Service Guidelines

Deafblindness: Educational Service Guidelines offers state and local education agencies a framework from which meaningful, appropriate programming for students who are deafblind can be developed. These guidelines identify the knowledge and skills educators need to assist their students who are deafblind reach their full potential and become successful, contributing members of our society.

Click on the following Chapters and Appendixes to jump to a description of the section.



Perkins School for the Blind

- Information on vision impairment
- Training opportunities (webcasts, library)
- Outreach services for students
- Families have attended evaluations
- Summer programs

Perkins Webcasts

- **CHARGE Syndrome: An Overview**

By Pam Ryan

In this webcast, Pamela Ryan, Perkins School Psychologist, offers an overview of the characteristic features of CHARGE Syndrome and discusses the very diverse ways these features may manifest themselves in children. She talks about some of the early medical complications that many children face and how these issues affect development and learning.

- **CHARGE Syndrome: Teaching Strategies for Children**

By Sharon Stelzer

Sharon Stelzer, a long term teacher in the Perkins Deafblind Program, discusses the impact of CHARGE Syndrome upon the student, and strategies a teacher can implement to create a good learning environment. Establishing schedules and structure as well as offering the student opportunities to make choices are stressed. Sharon also talks about the benefits of helping students with CHARGE Syndrome learn the art of negotiations.

- **CHARGE Syndrome: The Impact on Communication and Learning**

By Martha Majors

This very insightful webcast explains the physical, sensory and neurological issues shared by many children with CHARGE and how these issues can affect their success in school. Martha Majors, who has served many children with CHARGE in the Deafblind Program at Perkins, offers guidance for educators in developing an effective educational program that will improve the emotional wellbeing and success in learning for students with this syndrome.

Perkins Webcasts

- **Conversations: A Personal Reflection About Deafblindness**
By Barbara Miles In this webcast, Barbara Miles, a well-know as an author and lecturer, discusses her approach to engaging in conversations with students who are deafblind. She encourages people to think of how they converse with their friends and try to replicate the elements of those successful interactions in a way that is accessible to a child with limited vision and hearing. For example usually people initiate a conversation because the other person expresses a willingness to talk, through a smile or some other cue. Miles offers alternative strategies for making that connection when the person with whom you want to converse can neither see or hear you.
- **The Communication Portfolio**
By Susan DeCaluwe
In this webcast, Susan DeCaluwe discusses the development of the Communication Portfolio for learners with deafblindness and multiple disabilities. This tool, that is jointly developed by family members and professionals, creates a common and very personalized view of the learner's communication skills, abilities and challenges across all environments.

Perkins Webcasts

- **[Creating Vocational Portfolios for Students with Significant Disabilities](#)**
By Mary Zatta
[School-to-Work](#) helps educators to create meaningful vocational experiences for their students with significant disabilities and to develop vocational portfolios, essential tools as students transition to adult life. The book [School to Work](#), is currently available in the Perkins store.
- **[Early Literacy for Students with Multiple Disabilities or Deafblindness](#)**
By Deirdre Leech
Students with multiple disabilities, including deafblindness face many learning challenges. They do not learn literacy in typical ways. Often they do not have exposure to books and literacy based materials. Children with hearing loss may not have heard stories read aloud, and may not have used books on tape. The goal for these students is to maximize access using specialized formats.
- **[Love: Challenges of Raising a Child with Disabilities](#)**
By Jane Bernstein
Jane Bernstein, a parent and author of "Loving Rachel" and "Rachel in the World" - books which look at life with her daughter who has developmental disabilities was the keynote speaker at the 26th New England Regional Seminar for Children with Visual Impairments and Their Families (birth-7 years of age). This webcast is a tape of her keynote presentation.

General Findings

- Many children with multiple disabilities do not demonstrate intentional prelinguistic communication and object use
- Parents and teachers often do not “talk” to their child any more or any differently “after” the child receives the implant than before
- Many of the children with multiple disabilities are dropped from “therapy” if they do not make sufficient progress
- Some children have lost skills at 2nd or 3rd assessment

General Findings

- Parents and teachers are often not taught effective strategies to use with their children in natural routines and activities
- Part C service providers often teach isolated skills without working as a team to assist the parents to facilitate receptive and expressive communication forms and functions

Thank you

- Steppingstones of Technology Innovation
- Office of Special Education Programs
(#H327A0800045)

- Families who participated



 Cincinnati
Children's

change the outcome[®]