Language Outcomes of Young Children with Cochlear Implants and Additional Disabilities

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Cincinnati Children's

Objectives

- To quantify the post-implant language skills among deaf children with developmental disabilities who have received cochlear implants and evaluate the differences in these skills compared to hearing children with the same degree of disability

- To consider the implications for serving this population of children in early intervention systems

Rationale

- The prevalence of developmental disabilities among young children with hearing loss is estimated between 30-40% (GRI, Roberts)
  - 40% in our own CI population (Wiley)
  - 23% in Ohio EI medically complex

- The impact of dual disabilities has been theorized to have _more than an additive (multiplicative?) impact_ on child development
  - Studies have yet to quantify this impact

Deaf/hoh and Additional Disabilities

- This multiplicative effect on development may change how we provide interventions
  - Goal setting
    - How to prioritize goals for all of a child's needs
    - How do other issues impact processing of auditory skills
    - How do other issues impact ability to use hands for signing
  - Adapting educational techniques
  - Medical complexities and illnesses
  - Transition to school settings

CIs and Additional Disabilities

- Many studies are simply case series, descriptive, retrospective

- Comparison groups are rare
  - usually consist of typically developing deaf children with implants

- Testing batteries may not be appropriate to all children in the group

- Vast heterogeneity in children with additional disabilities

- Studied outcomes may not be appropriate for all children

CI and Cognitive Delays

- Lower levels of <u>speech perception</u> and slower progress (n=20 cog/motor delays). 60% not achieving <u>open-set word recognition</u> (Pyman et al, 2000)

- Most with mild cog delay (n=14) achieve <u>open-set recognition</u> of familiar words (Dettman et al, 04)
  - Highly variable with more significant delays (n=8)

CI and Cognitive Delays

- Slower rate of improvement in <u>sentence recognition</u> and lower language quotients among mild cog delays (n=19) (Holt & Kirk, 05)

- Children with delays (n=11) may progress in <u>speech perception/production</u>, but not like typically developing children (Edwards, '06)
  - Children with significant delays showed almost no progress in either domain

CI and Cognitive Delays

- All studies compared children with cognitive delays and CIs to *typically developing children* with CIs

- Thus, results are not surprising. Children with cognitive delays would likely have slower rates of outcome progression than typically developing peers

Autism Spectrum Disorder

- Donaldson et al 2004
  - Descriptive study of 7 children with autism spectrum and an implant
  - Most outcome data from parent report measures
  - Wide variability in outcomes
  - Improvements in <u>auditory skill development</u> and/or <u>receptive vocabulary</u> for 3 of the 4 that could complete these measures

Deaf-Blind

- Young et al 1995 (n=4 US Type I)
- Saeed et al 1998 (n=2)
- El-Kashlan et al 2001 (n=2)
- Damen et al 2006 (n=7, US Type I)
- Pennings et al 2006 (n=10, US Type I)

- In general, children with Usher Syndrome Type I would be expected to have average cognitive abilities
- Important to consider full auditory skill acquisition rather than anticipating being able to rely on lip-reading

Mixed Disabilities

- Studies report variability in improvements (types and amount), much is anecdotal

- Improvements in speech and/or word recognition seen in **10-70%**
  - Hamzavi et al ('00) 2/10; Waltzman et al ('00) 12/29; Vlahovic & Sinija ('04) 4/4 "good" speech perception;
  - Winter et al ('04) only 1/10 word recognition,
  - Berrettini et al ('08) 12/23, excellent speech perception
  - Nikolopolous et al ('08) 47/67 developed connected intelligible speech 5 years post implant

Mixed Disabilities

- Improvements in detection, pattern perception
  - Winter et al 10 children detect LING-6, 8/10 pattern perception

- Auditory skills
  - Daneshi 07 Improved auditory perception in all 55 patients
    - Difficulties testing due to limited vocabulary, attention span, etc
  - Wiley 08 all 14 children had improvement auditory skill development
    - <u>Nonverbal cognitive abilities</u> had great impact in rate of progress, more so than the disability

Mixed Disabilities

- <u>Qualitative/perceived benefits</u> (Wiley, 05 [n=16]; Berrettini, 08 [n=23])
    - Parent's perception positive, would make same decision
    - 100% improvement in environmental awareness
    - More likely to communicate wants and/or needs

- <u>Anecdotal</u> reports of
    - general connectedness to the environment (Walzman,'00)
    - qualitative benefit of increased auditory awareness, improved motor skills, reduced hyperactivity (Hamzavi '00)
    - increased self-sufficiency in all, results were primarily descriptive ("satisfying results" for autism, more curious and actively participating in everyday interactions, improvements in balance (Filipo, '04)

Study Research Question

- We know that children with additional developmental disabilities have deficits in a variety of outcomes
  - What is the impact of HL itself in this population?

- What are the language outcomes associated with receiving a cochlear implant?
  - Pre to post changes
  - Compared to "peers"

Current Study

- **Aim 1:** To test whether deaf children with additional disabilities have improved language skills after receiving a CI.

- **Aim 2**: To test whether language skills of deaf children with additional disabilities who receive CIs are lower than hearing children with similar disabilities.

- **Aim 3:** To test the whether pre-CI developmental quotients of children with disabilities are predictive of language skills post-CI.

Developmental Disabilities and CI

- One of the most difficult aspects of studying outcomes among children with additional disabilities is the choice of an <u>appropriate control group</u>

- ***The language skill set in children should be appropriate/commensurate with their developmental level***

Design Methods

- Cross-sectional design

- Children ages 2-8

- One time language evaluation using the PLS-4

- Matched to children with hearing according to <u>age and cognitive abilities</u>

- Parallel longitudinal study
  - Includes original cohort for 1 year post-study follow up
  - Prospective cohort pre-CI
  - Language and functional assessment

Statistical Methods

- Aim 1: Language differences between groups tested using Wilcoxon Sign Rank test (matched-pairs analysis) (Multiple regression analysis is ongoing)

- Aim 2: Pre-Post CI differences tested using Wilcoxon Sign Rank Test and multiple regression

Main Outcome

- Receptive and Expressive Language Quotients on the PLS-4
  - Defined as the language age obtained on the PLS-4 at the time of the study divided by the chronologic age of the child at the time of the study

- Children were tested with and without sign support if sign language was a communication strategy of the child

Developmental Assessment

- All CI children received assessment by the same developmental pediatrician
  - Occurred prior to 3 years of age

- Revised Gesell Developmental Schedules
  - Provides age equivalents
    - Nonverbal cognitive (performance), gross motor, fine motor, personal social, language (not used for study)
  - Developmental quotients calculated by dividing age equivalent for nonverbal cognitive by chronologic age at time of testing

Other Factors Collected

- Child specific factors
  - Ages at HL diagnosis and CI, Duration with implant, School setting and duration in school, Types of interventions/therapies and intensity

- Family specific factors
  - Highest education, income, other siblings in household, insurance

To Date

- 21 subjects with enrolled in CI group
  - 2 no developmental information
  - 1 subject cognitively typically developing

- 15 currently matched controls
  - Enrolling 3 additional controls for study completion

CI Subjects

HL Etiology	
CMV	5
CHARGE syndrome	4
Genetic or EVA	3
Non-CMV infectious	3
Other	4

Developmental diagnosis	
Cognitive delay	5
CHARGE syndrome	4
Cerebral palsy	4
Multiple	2
Other	2

	CI (n=15)	Controls (n=15)	p
Age of child*	52 (28-81)	65 (31-81)	0.05
Non-Verbal Cog Quotient	50 (33-92)	54 (30-89)	0.37
Gender – male	53%	80%	0.16
Insurance type (private only)	42%	31%	0.71
Mat educ (beyond HS)	77%	69%	0.66
Income < $40,000	42%	54%	0.48
Receiving sp/lang therapy	85%	69%	0.27
# different therapies	**3 (1-5)**	**2 (0-5)**	**0.02**
Total hours/week in therapy	**3 (0-26)**	**0 (0-7)**	**0.04**
Total hrs/wk in sp therapy	**1 (0-12.5)**	**0.5 (0-2.5)**	**0.09**
Age at CI	21(13.5-51)	---	
Duration since CI	25 (10-68)	---	
Age at HL identification	3 (0-25)	---	
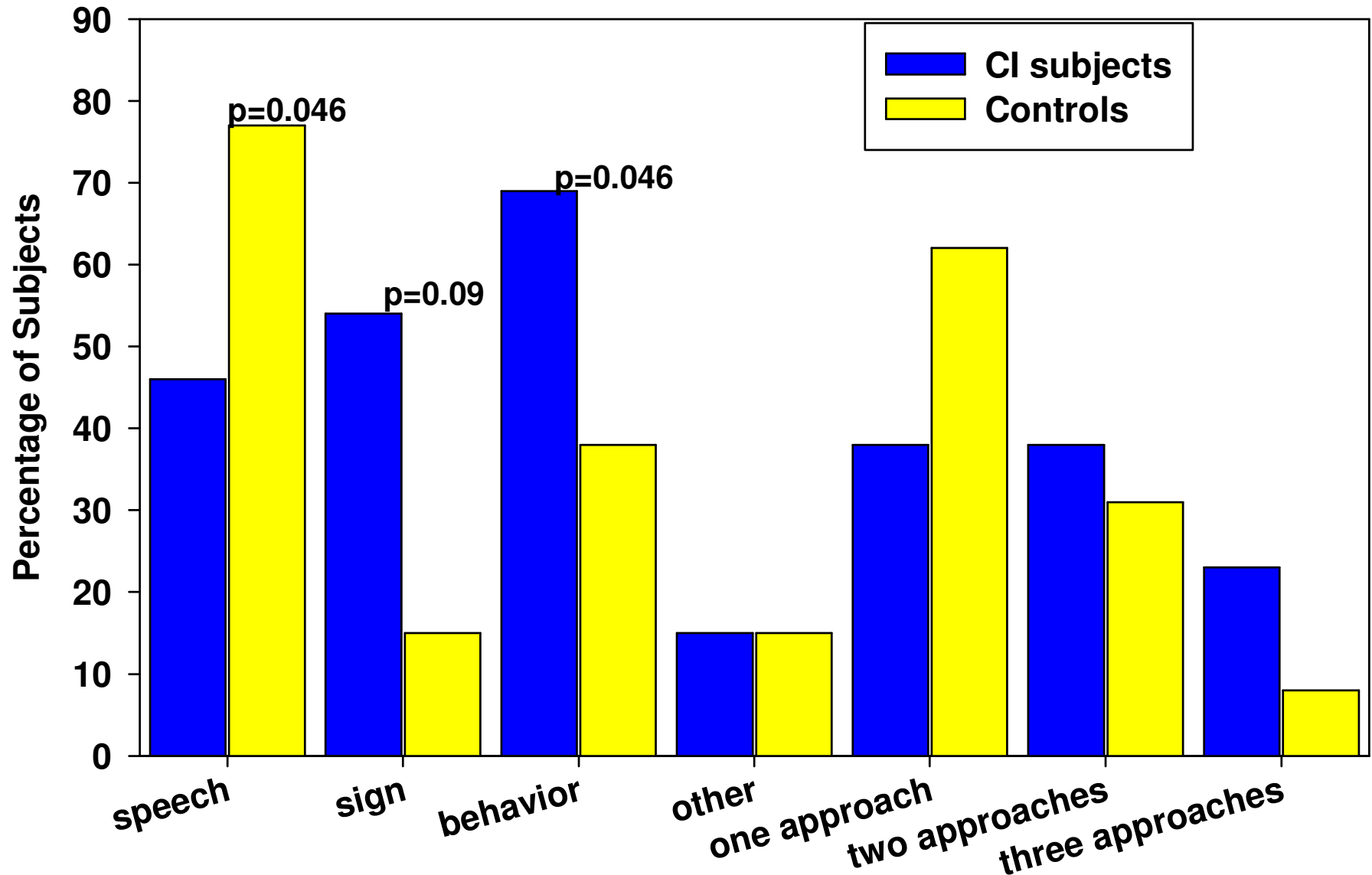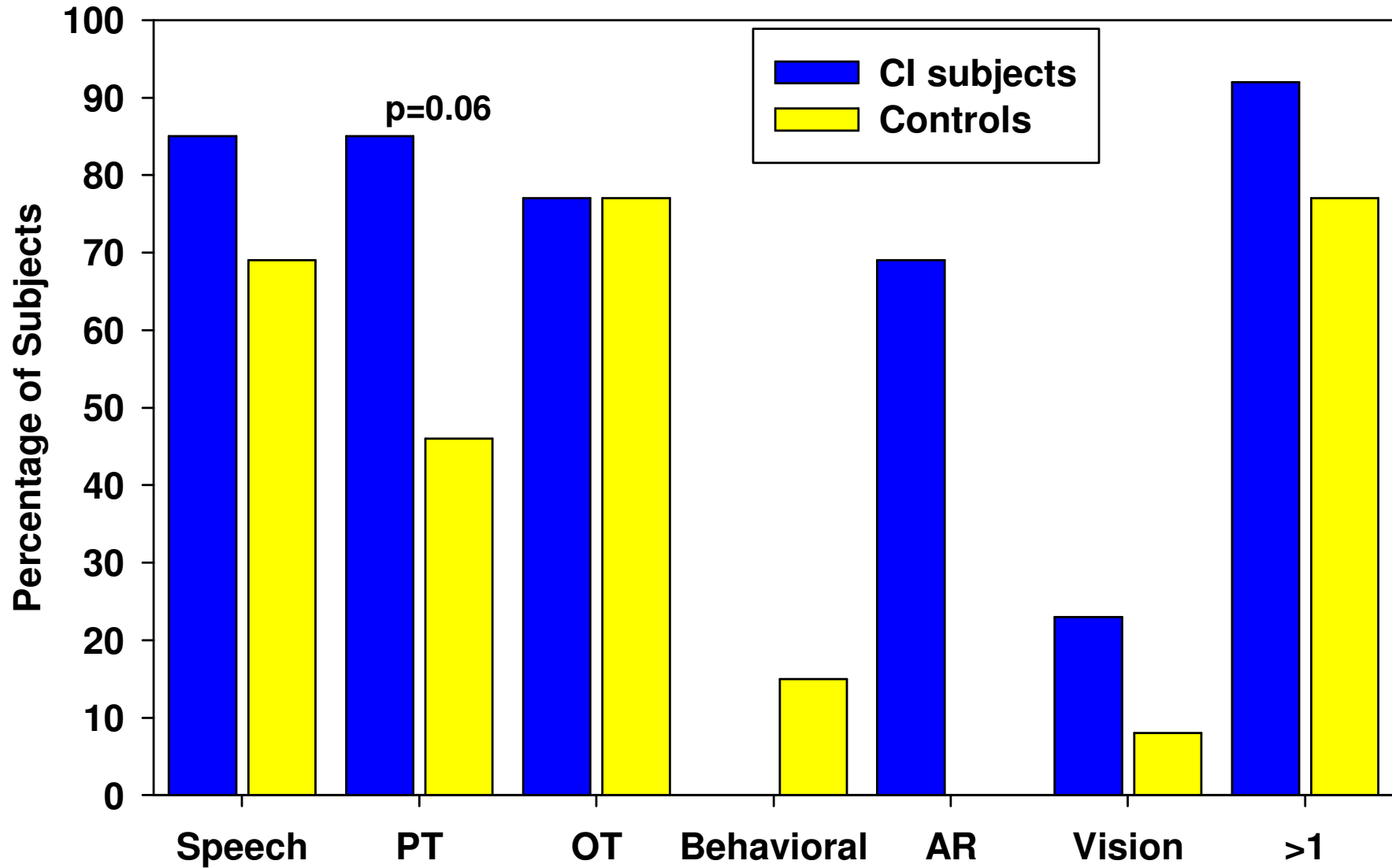
Types of Interventions
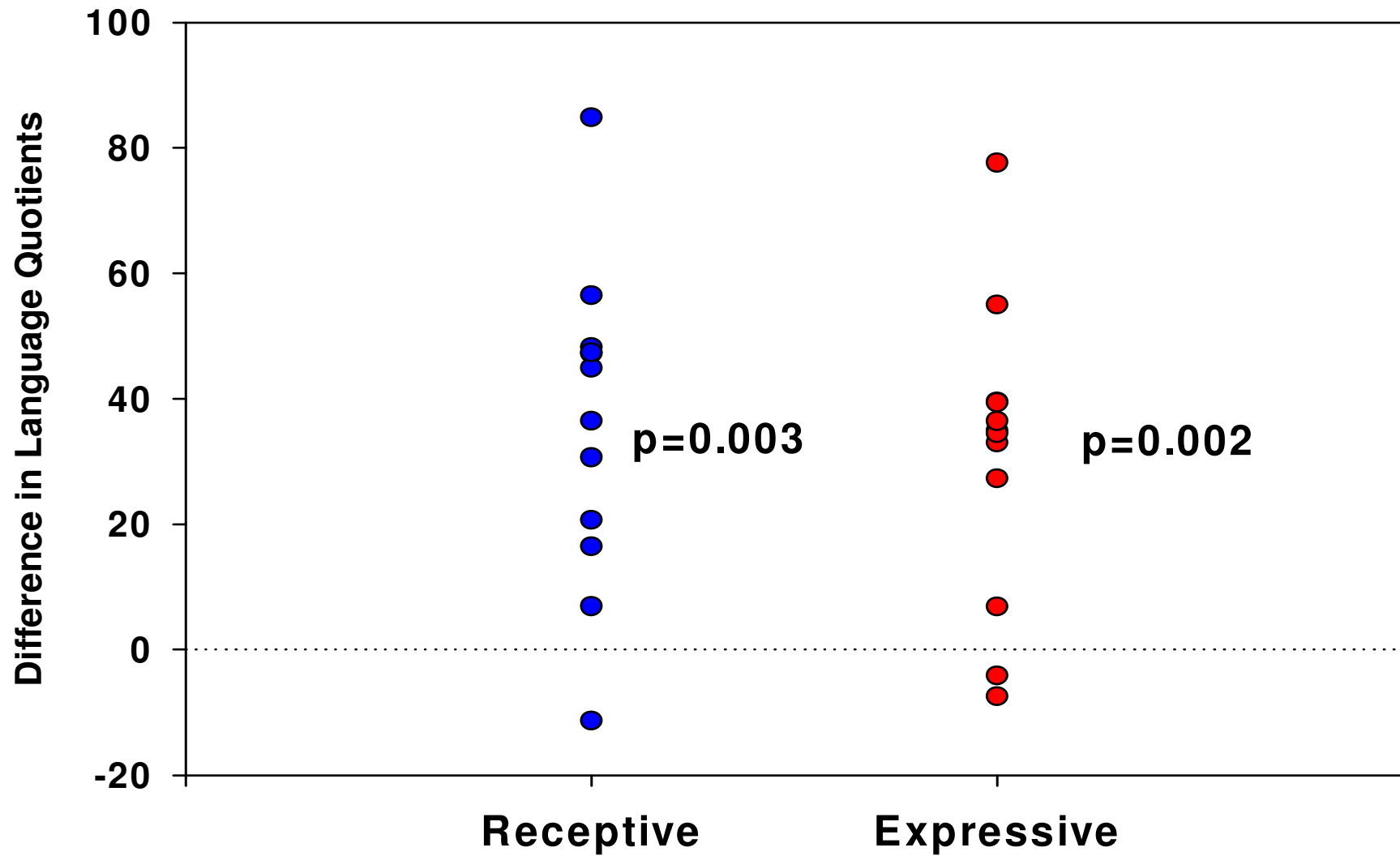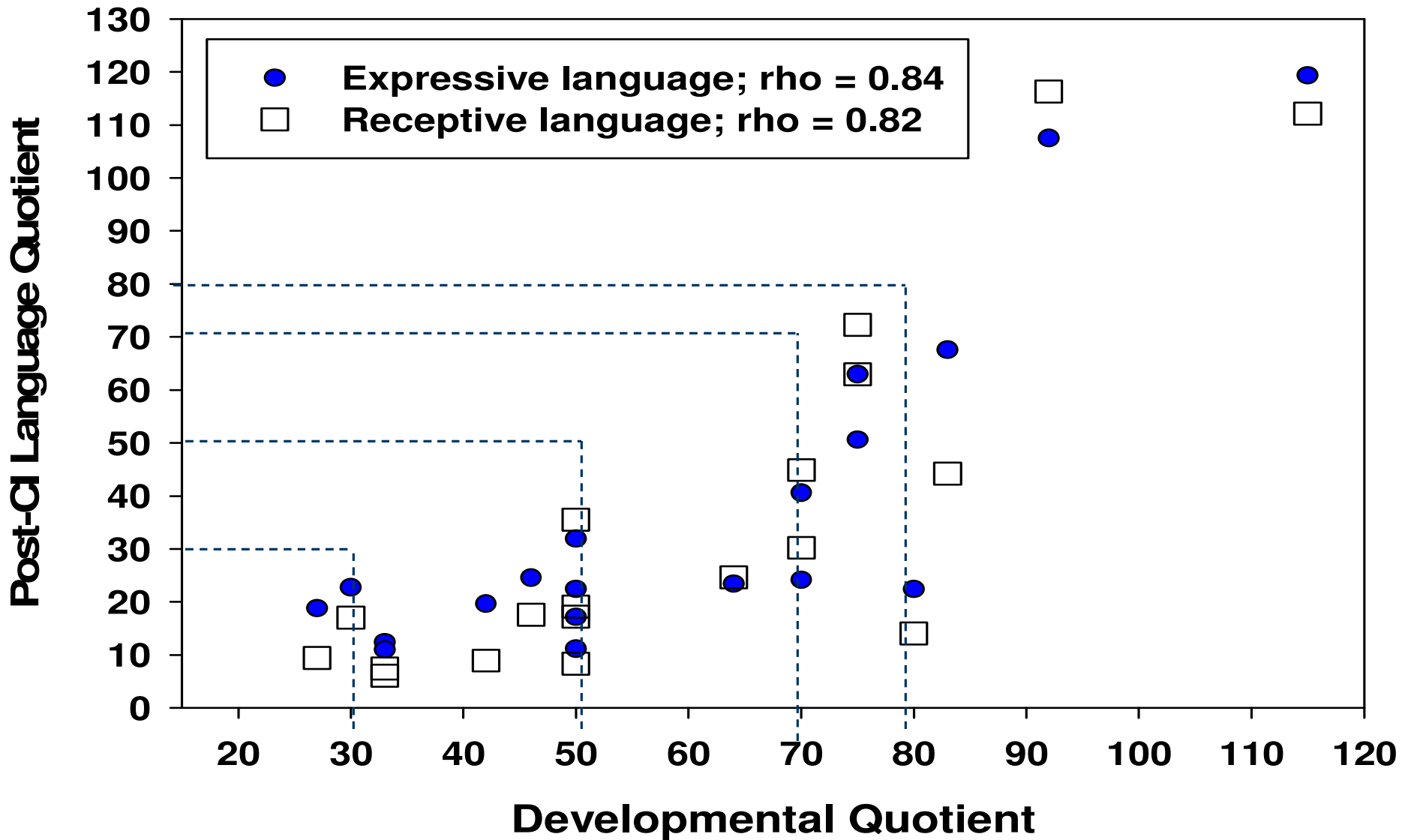
CI: DQ and current LQ

Controls: DQ and current LQ
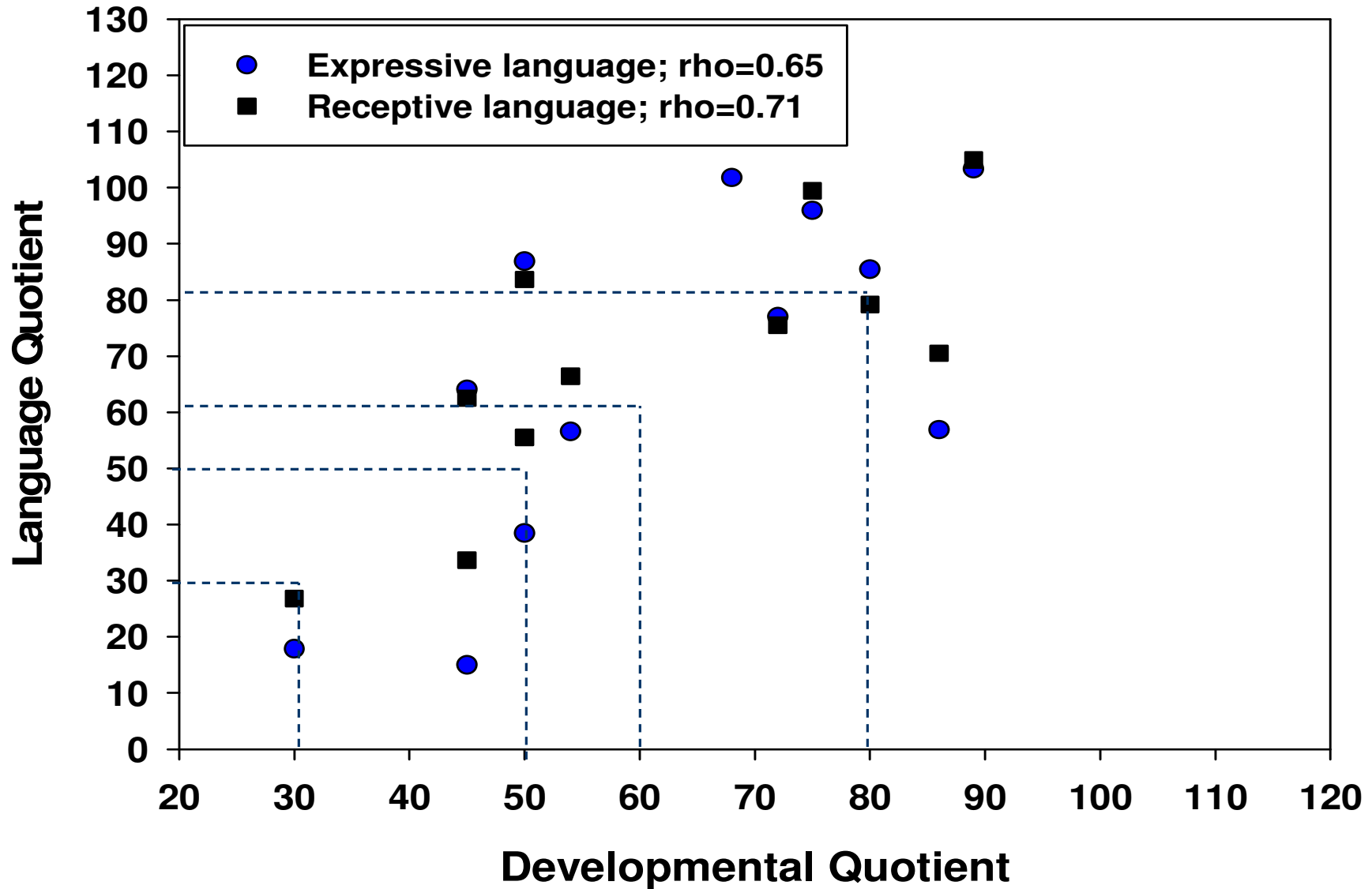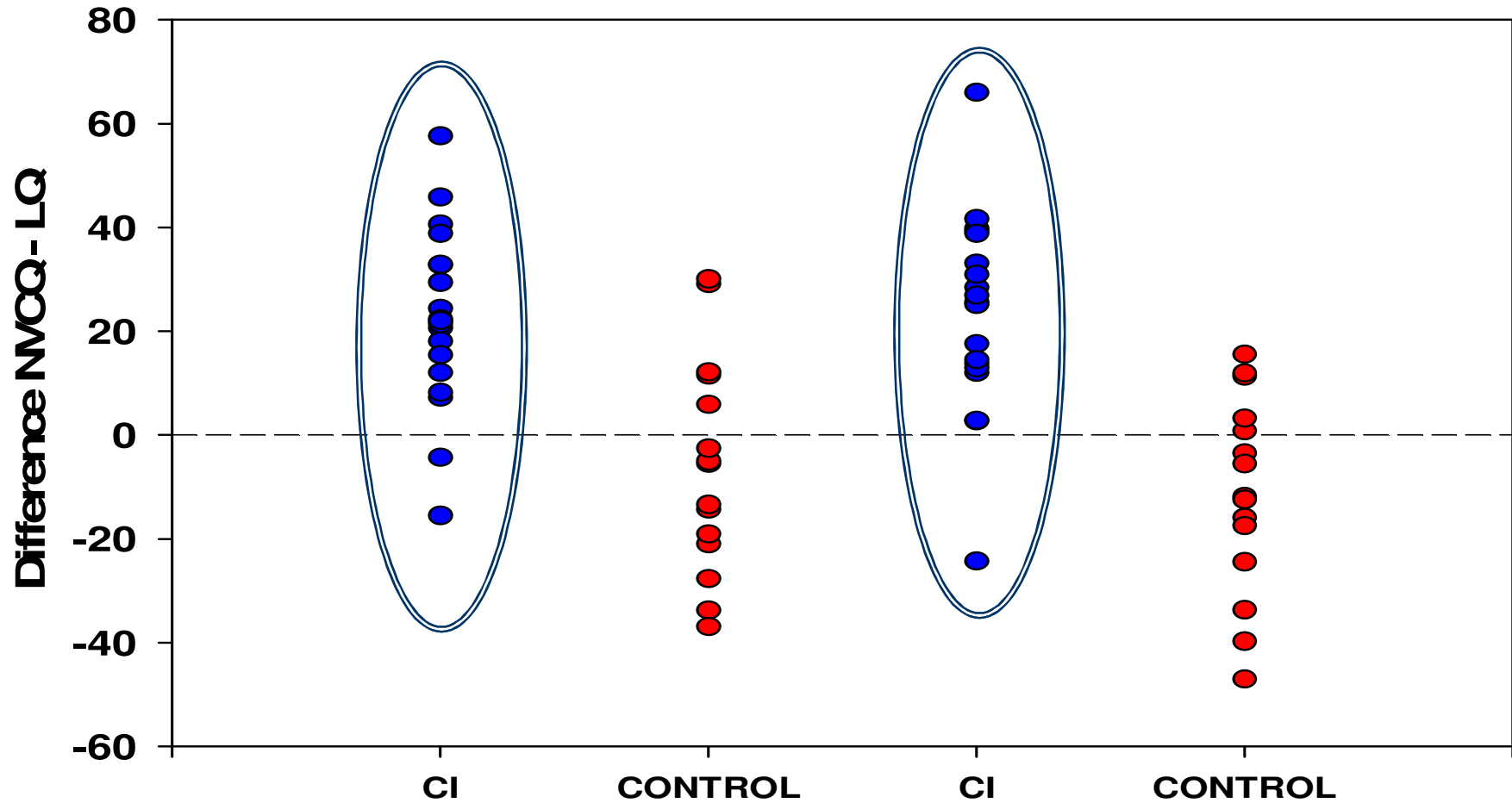
Pre-Post CI Language Development

- 14 subjects included who had complete pre-CI language testing available

- Rosetti Infant Toddler Language Scale used as part of clinical protocol for 0-3 yrs
  – Provides language age equivalent

- Language quotients determined by dividing language age equivalent by chronologic age of child

Pre-Post Language Methods

- Correlations conducted regarding change in language (language age and language quotient)

- Absolute changes between pre and post results

- Multiple regression models constructed to determine independent predictors of language
  - Outcome LQs at time of study, controlling for pre-CI LQs and duration of implant
  - Determine what proportion of the variance in the LQ was due to specific predictors (i.e. cognition)
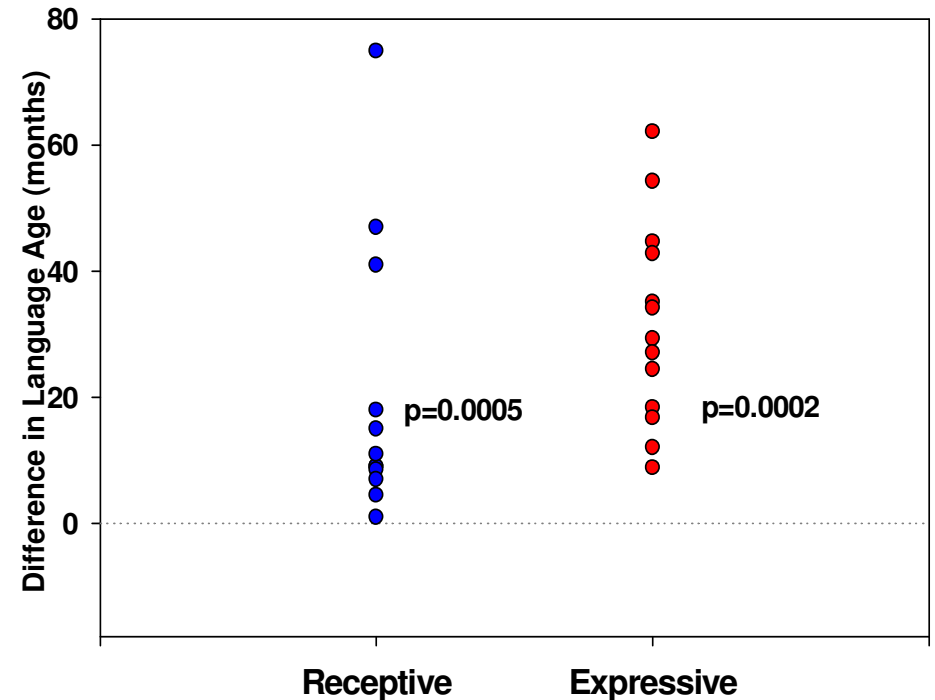
Pre-post CI Differences

Language Quotient
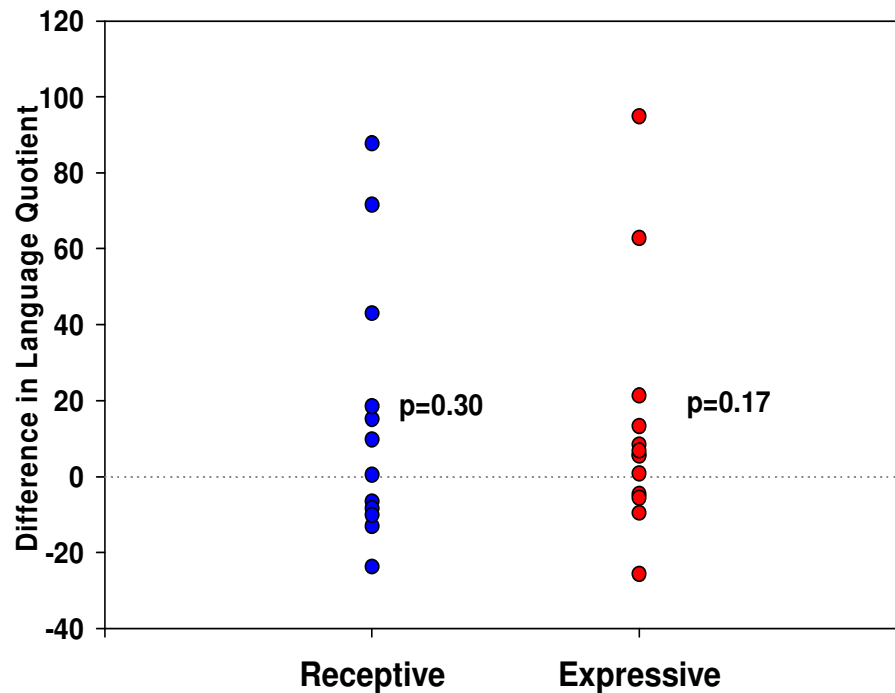
Language Age

•No significant changes seen with language quotient, but everyone made some language process according to increase in language age

•Increase in LQ means that children are closing the language gap between chronologic and language age

Pre-Post Language Results

- No association/correlation found between <u>change</u> in LQs and the following:
  - Age of implant, duration of implant, number of issues, total number of therapies, hours in language therapy

- No association regarding gender, income, insurance status

- Cognitive abilities significantly correlated ($p<.05$) to change in LQ and change in language age

Rate of Language Progress and Cognition

- Receptive
- Expressive

Y-axis: Increase in Language Age per Month
X-axis: Nonverbal Cognitive Quotient
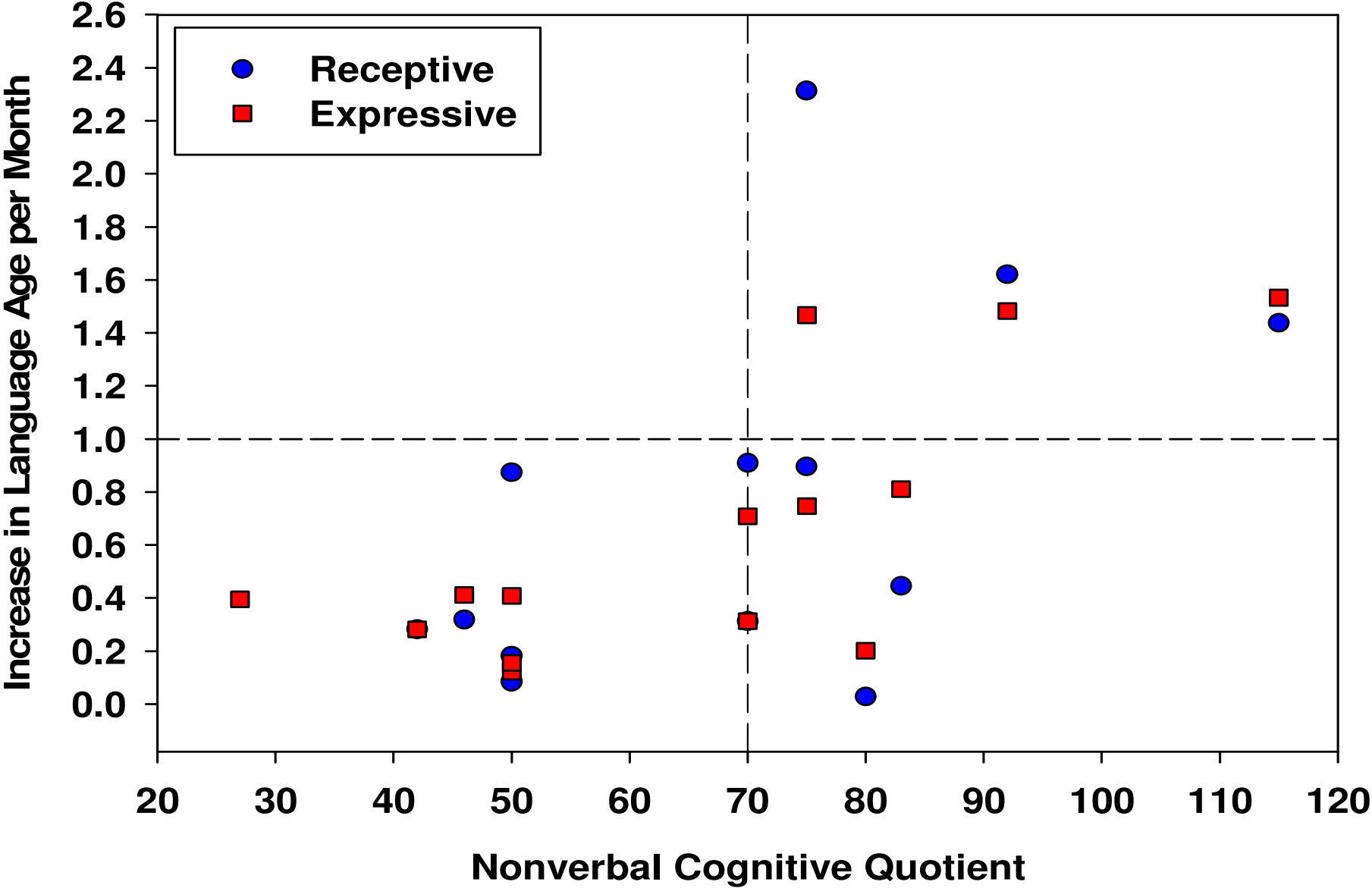
Multiple Regression Results

Predictor	Amount of Variance explained (R^2)	P-value
NVCQ	64%	<.0001
Age at diagnosis	16%	<.0001
Parental education	12.6%	0.001
Duration with CI	5%	0.002
Pre-CI LQ	0.4%	0.53
98% of variance explained by 5 factors		

Other Interesting Findings

- In medically complex children, therapeutic focus may change based on developmental needs (i.e. feeding vs language)

- In a subset of children, adding sign support increased their receptive and expressive language levels

- Clinically, children CI and co-existing disabilities potentially need more monitoring of progress, and consideration of adaptive/augmentative strategies

Two anecdotes

- Child with CHARGE, trach, G-tube, etc.
  - Had speech therapy for feeding, however was still aspirating and goal for feeding was unlikely currently
  - Being in the study, prompted family to begin to use pictures for communication
  - Shifted speech therapy to focus on communication

- Child with severe cerebral palsy
  - Following study, family looked into eye-gaze systems
  - Augmentative communication evaluation is occurring (had been evaluated previously, but center had thought she was "too low")

Impact for Early Intervention?

- Can we start to close this gap?
- Should we be taking a **different** therapeutic approach for these children?
- How often should we **monitor** progress and change strategies?
- Counseling on **appropriate expectations**, goal setting (goals not too high, not too low)
- High need for **collaboration** with other therapists (behavioral, OT, PT, ABA, etc)
- **INSITE** training

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change the outcome

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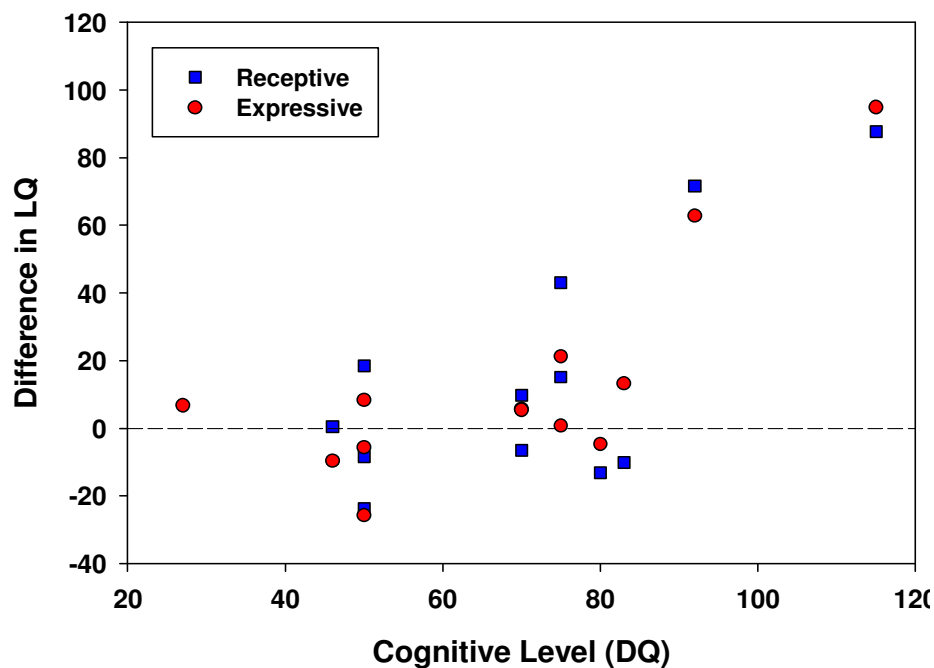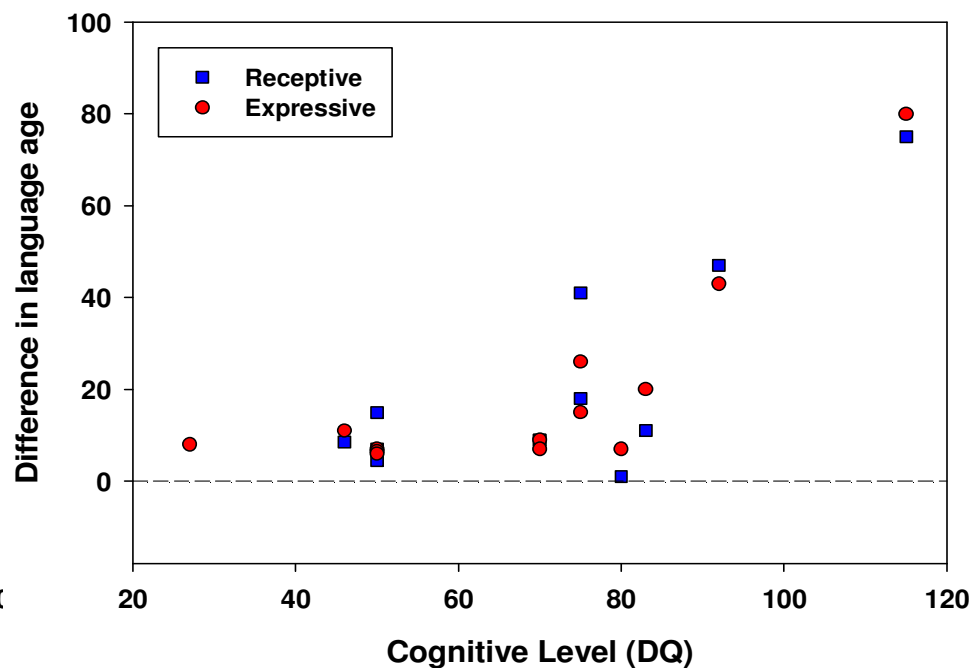
Pre-Post CI Language Differences
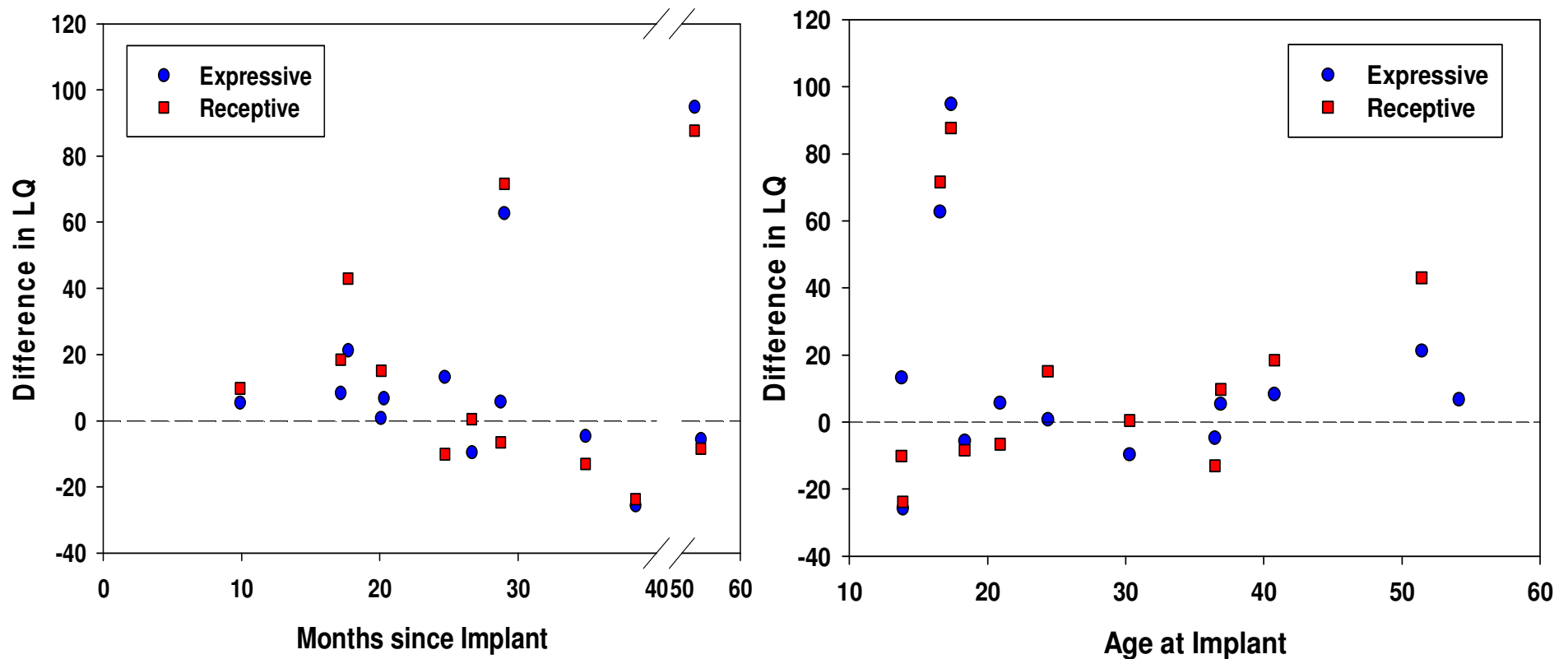
No correlation with age of CI or experience, even after controlling for cognitive abilities

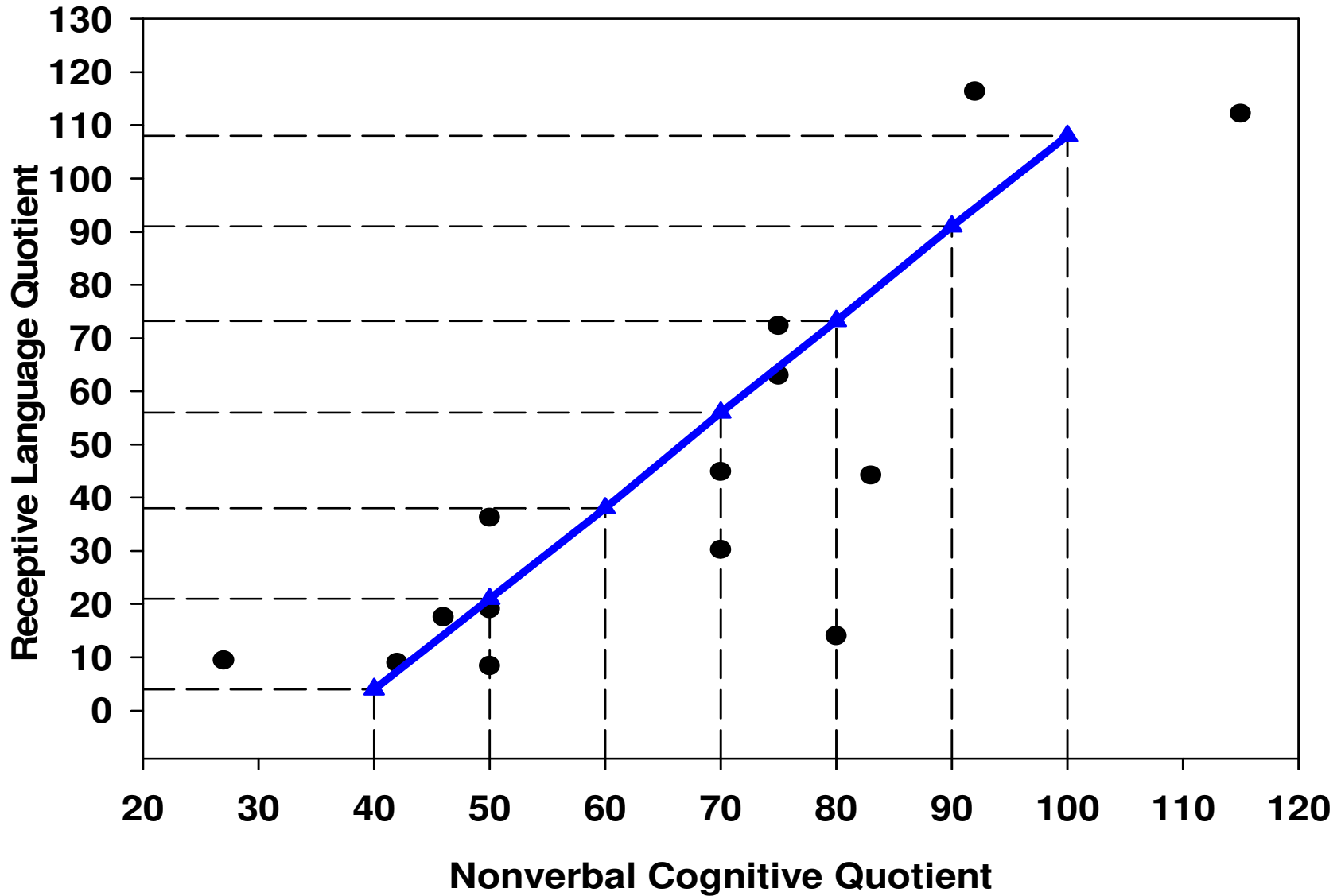# Pre-Post CI Changes in Language by Cognitive level

Cognitive abilities appears to be significantly correlated (p<0.05) the change in both LQ and age

Multiple Regression Results

Difference in LQ between sign and auditory language testing