

Early Diagnosis of Cerebral Palsy to Promote Engagement, Plasticity and Family Well-Being

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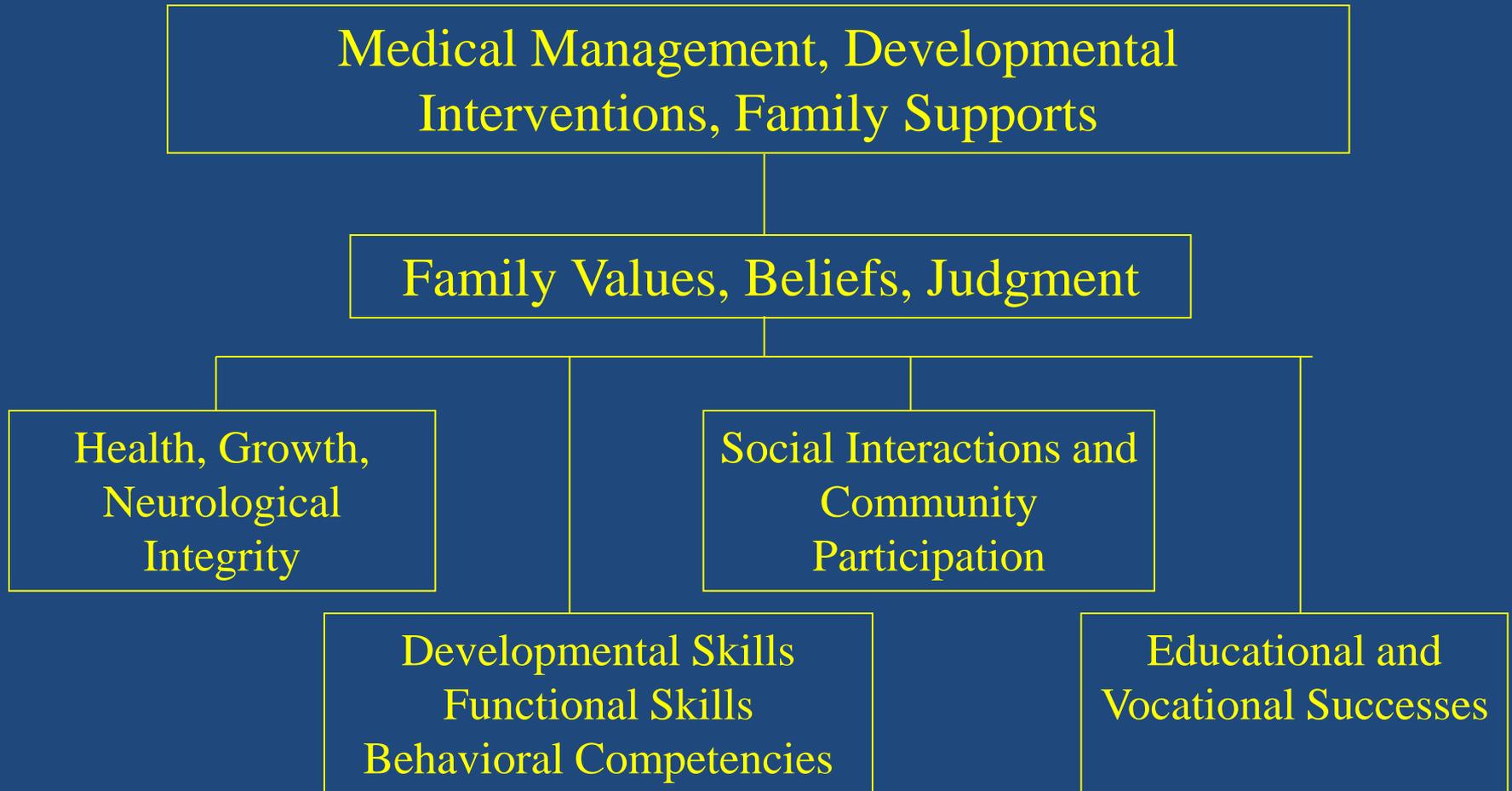
Neuroprotection in VP and EP

- Every week matters
- Maternal corticosteroids
- Surfactant replacement
- Avoid pneumothorax
- Prevent sepsis
- Optimize nutrition
- Timely and appropriate developmental inputs.

Key Questions

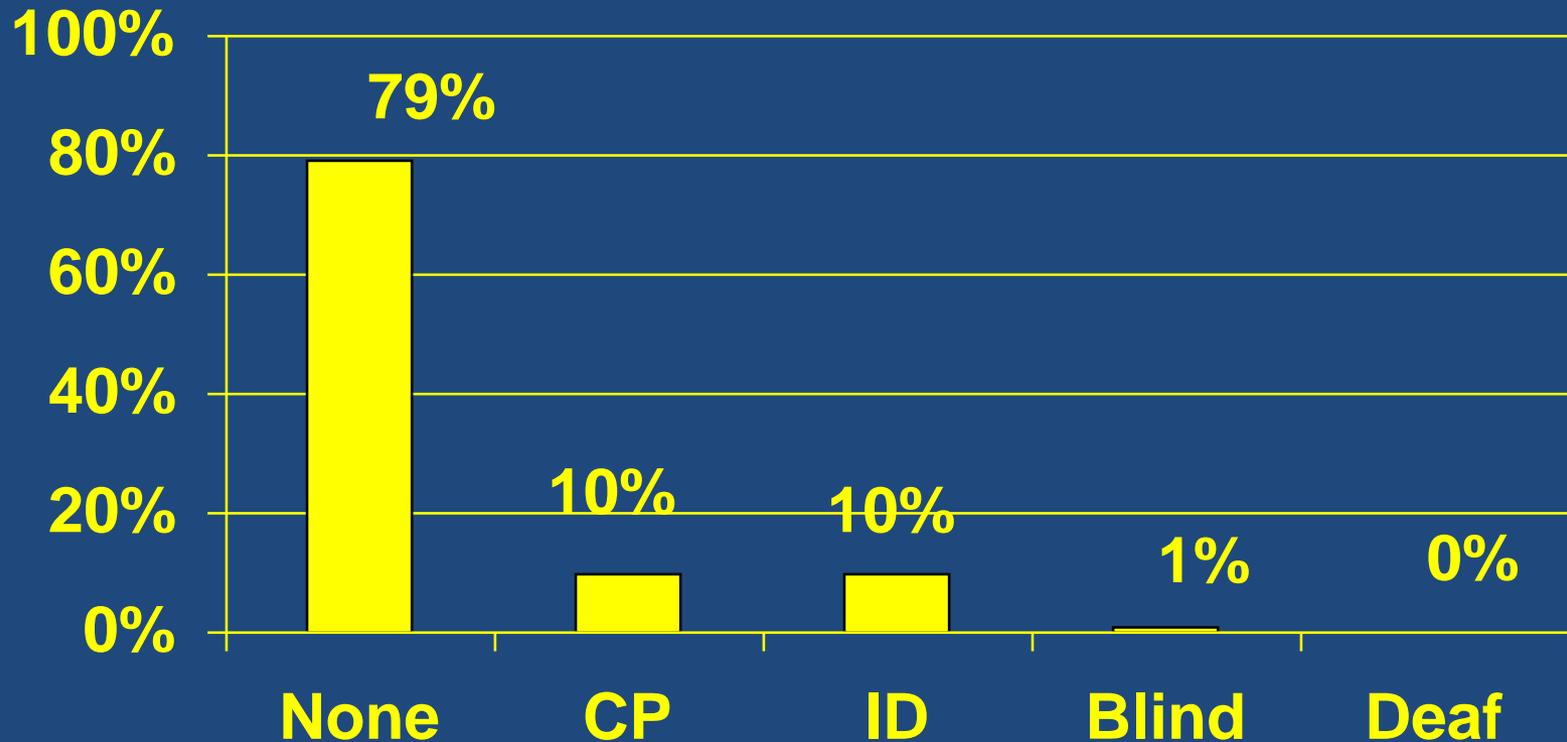
- What happens to premature babies?
- If they have disability, what is the impact on child functioning ?
- What are the myths of outcomes after prematurity?
- What are the myths of outcomes after disability?
- What are the sources of information that can inform health, educational and community professionals?
- How can we find ethical web sites?
- How can we find information that is enabling?

Measuring Quality of Life in Child Neurodisability



-Adapted from Spilker

Neurodevelopmental Disabilities



Neurodevelopmental morbidity among survivors (N=149). CP = cerebral palsy; ID = intellectual disability

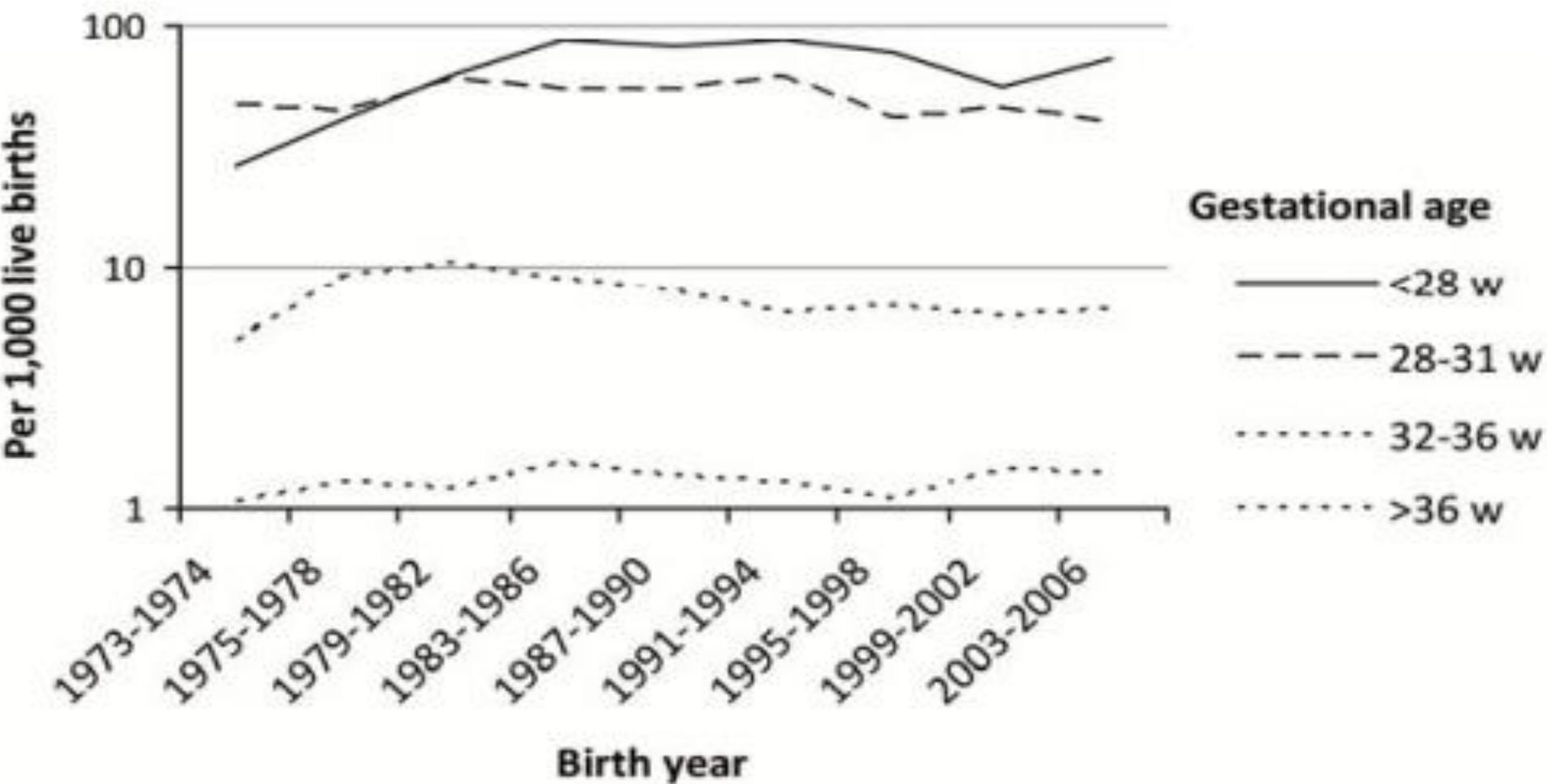
Kwong, et al. Pediatrics 1985; 76: 585-92 and 1986;78: 767-74

Functional Status at Kindergarten Entry

Walks 150 Feet	97%
Talks in Sentences	97%
Toilets Self	96%
Self-Care/ ADL	96%

Functional Status and Neurodevelopmental Disability

Walks 150 Feet	87%
Talks in Sentences	84%
Toilets Self	81%
Self-Care/ ADL	81%
Understands Requests	94%



Acta Paediatr. 2014 Feb 27. doi: 10.1111/apa.12614. [Epub ahead of print]

The panorama of cerebral palsy in Sweden. XI. Changing patterns in the birth-year period 2003-2006.

Himmelmann K1, Uvebrant P.

Epidemiology of Cerebral Palsy I

- Term: 1 per 1000
- Preterm 32-36 wks: 10 per 1000
- Very Preterm 28-32 wks: 70 per 1000
- Extreme preterm 23-27 wks: 100-160/1000

Early, accurate diagnosis and early
intervention in cerebral palsy:
Advances in diagnosis and treatment

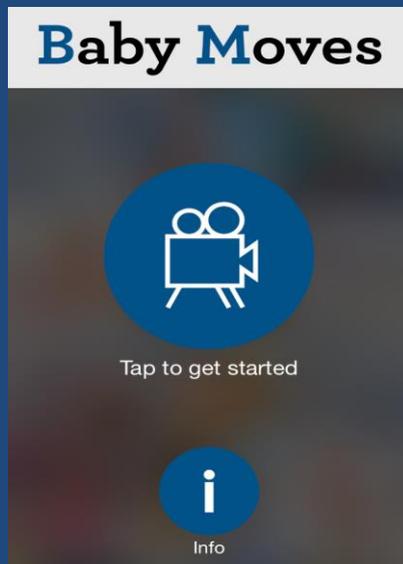
Novak I, Morgan C, Adde L et al.

JAMA Pediatrics, September 2017

	Sensitivity	Specificity
General movement assessment	98%	91%
Cranial ultrasound	74%	92%
MRI	86%	89-97%
Neurologic examination	88%	87%

Baby Moves

- 3 months CGA
- Can evaluate for CP, risk of other adverse neurodevelopmental outcomes



How To

Making a 3 minute video of your baby

Your baby should be:

- Lying on his/her back
- Awake and moving
- Happy (not crying)
- Wearing singlet and nappy only

Tips:

- Take out dummy
- Take away toys
- Try not to get your baby's attention
- Keep hands and feet in view



What do we know?

- **Strong** evidence for early prediction of cerebral palsy
- GMA is a low cost tool that can be used in high and middle/low income countries
- Reassures worried families with children at high risk
- Directs resources to those at greatest need at time of high brain plasticity
- Potential as biomarker in clinical trials

Communication Matters

- Communication should be face-to-face, with both parents or caregivers, private, honest, jargon free, empathetic, tailored to the family
- Requires current written information, identification of strengths, invitations to ask questions, discussions of feelings, recommendations to use parent-to-parent supports, and arrangement for early interventions
- Remember parents worst fears include multiple neurodevelopmental challenges including neurodegeneration.
- Need to foster practice of self-assessment based on Reflection, Humility, Openmindedness, Partnership, and Engagement(R-HOPE).

Reflection

- Ability to look critically at one's attitudes and behaviors thru self-examination & discussion
- Clinicians may influence parents consciously or unconsciously
 - Awareness of one's own values and opinions, and the possibilities of biases that influence decision-making
 - Prognosis can be influenced by a clinician's unacknowledged emotional reactions
- **Requires** critically considering attitudes toward initial prognosis or eventual disability

Humility

- A modest attitude with respect to the evaluation of one's importance
- Much uncertainty exists about neurological prognostication, which calls for a careful and humble attitude
- Requires sensitivity to receptivity of parents to uncertainty consistent with variations in decision-making preferences
- **Reflects** the unpretentious open attitude (with honest self-disclosure of uncertainty and avoidance of arrogance) consistent with a patient-centered approach

Open-mindedness

- Receptiveness to medical outcomes which defy one's prognosis and willingness to fairly consider prognosis that differ from one's own
- **Physician** must acknowledge how age, experience, cultural and religious background, training, and geography modulate prognosis and attitudes about life-sustaining therapies.
- Open-mindedness is a desirable /attitude for clinicians in service of families in settings of uncertainty

Partnership

- Collaborative discussion about goals of care; reflected in communication and style of clinician with parents as intrinsic and important actors in the care team
- Large amount of evidence that there are differences in parent, child, and clinician ratings of quality of life
- Clinicians often more pessimistic
- Good practices use a personalized model of care based on parental needs without overstressing family

Engagement

- Commitment of clinician to step outside comfort zones in pursuit of child and family well-being using best scientific practices
- Information and good communication alleviate feelings of estrangement felt by parents in the NICU environment
- Engagement entails a broader view of neonatal outcomes and corresponding gestures on behalf of clinicians
- Engaged clinicians are committed to evidence-based medicine and outcomes of child and family well-being.

Conclusion

- OuR-HOPE approach presents desirable clinical practices which reflect attitudes and behaviors responsive to clinical situations to support ethical communication and decision-making
- Further discussion and research needed to support the integration of more adequate, evidence-informed practices in communication of neurological prognosis in the neonatal context

Case 1: A child with visual and motor delays

- A child is adopted from an Asian orphanage
- At 3 months he demonstrated nystagmus and saw pediatric ophthalmology and neurology
- At 6 months, he had sitting balance and used right hand for reaching and transfers.
- At 9 months, he could not quadruped crawl , he played peek a boo, and finger fed crackers.

Case 1 A child with schizencephaly and septo-optic dysplasia

- At 12 months, MRI revealed schizencephaly with partial agenesis of corpus callosum consistent with SOD
- He said mama, dada, milk, and ball.
- He released blocks into a cup, had object permanency, localized bell, and put pegs in.
- His neurological exam revealed left arm held in flexion with fisting, no left lateral prop or parachute, clasp knife hypertonus at elbow, and brisk reflexes with positive babinski.

Case 1 Counseling and CNS dysgenesis

- At age 2 years, he has had one year of constraint. He can use left arm to assist. He solves 10 piece puzzle, talks in sentences, has a vocabulary of >200 words, uses a spoon, removes socks.
- At age 5, he reads Dr. Seuss, keyboards one handed, jogs with his dad, and sometimes has urinary accidents.
- At age 8 he has a verbal and nonverbal IQ of 135, is an A student in all subjects, but needs visual supports.

Key Messages

- Cerebral Palsy is a disorder of movement and posture due to causal pathways impacting on developmental brain connectivity.
- There is a spectrum of oral, fine, and gross motor challenges in functioning.
- Important co-morbidities include vision, hearing, seizures, and feeding.
- All children with CP learn.
- All children with CP benefit from activity that promotes functioning, participation, and independence.
- Enriched environments matter and should not be delayed by bureaucratic mischief.
- Neuroprotection and Neuroplasticity require our-HOPE, enablement, and networks committed to maximizing resilience.

IHDP Interventions

- Program began at neonatal discharge
- All children received medical care
- All children had serial developmental and social assessments
- All control children with problems were referred for appropriate treatments

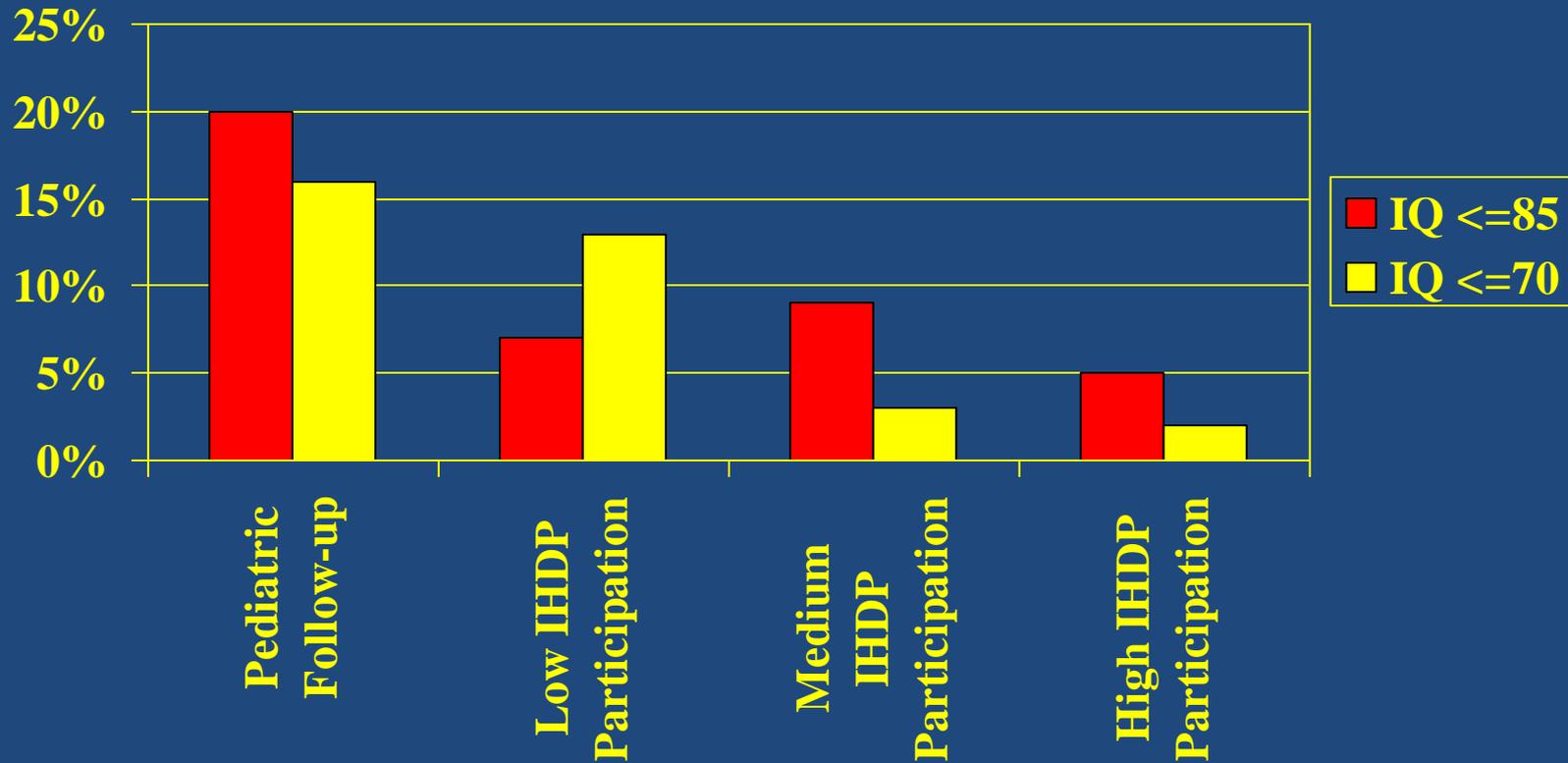
IHDP Experimental Intervention

- Weekly home visits in year 1
- Biweekly home visits in years 2 and 3
- Curriculum for relationship based learning and communication skills
- Curriculum for parent problem solving with child behavior and social stressors

IHDP Early Childhood Intervention

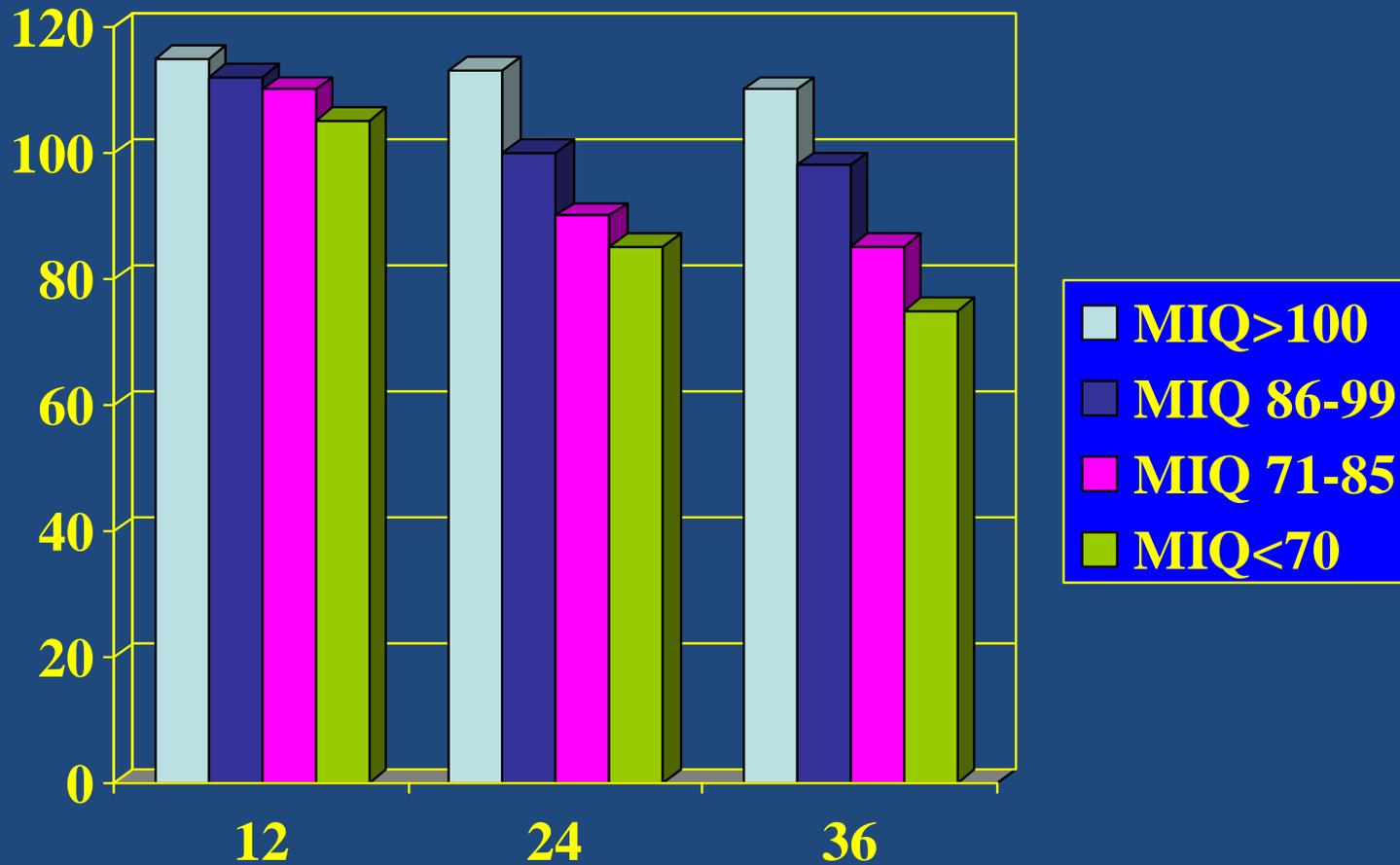
- Center based interventions 50 hrs per wk
- Staff ratio 1:3 at 12-24 months
- Staff ratio 1:4 at 24-36 months
- Teachers were college graduates
- Learning Games was the core curriculum

Follow-up (Control) and Intervention Participation Groups

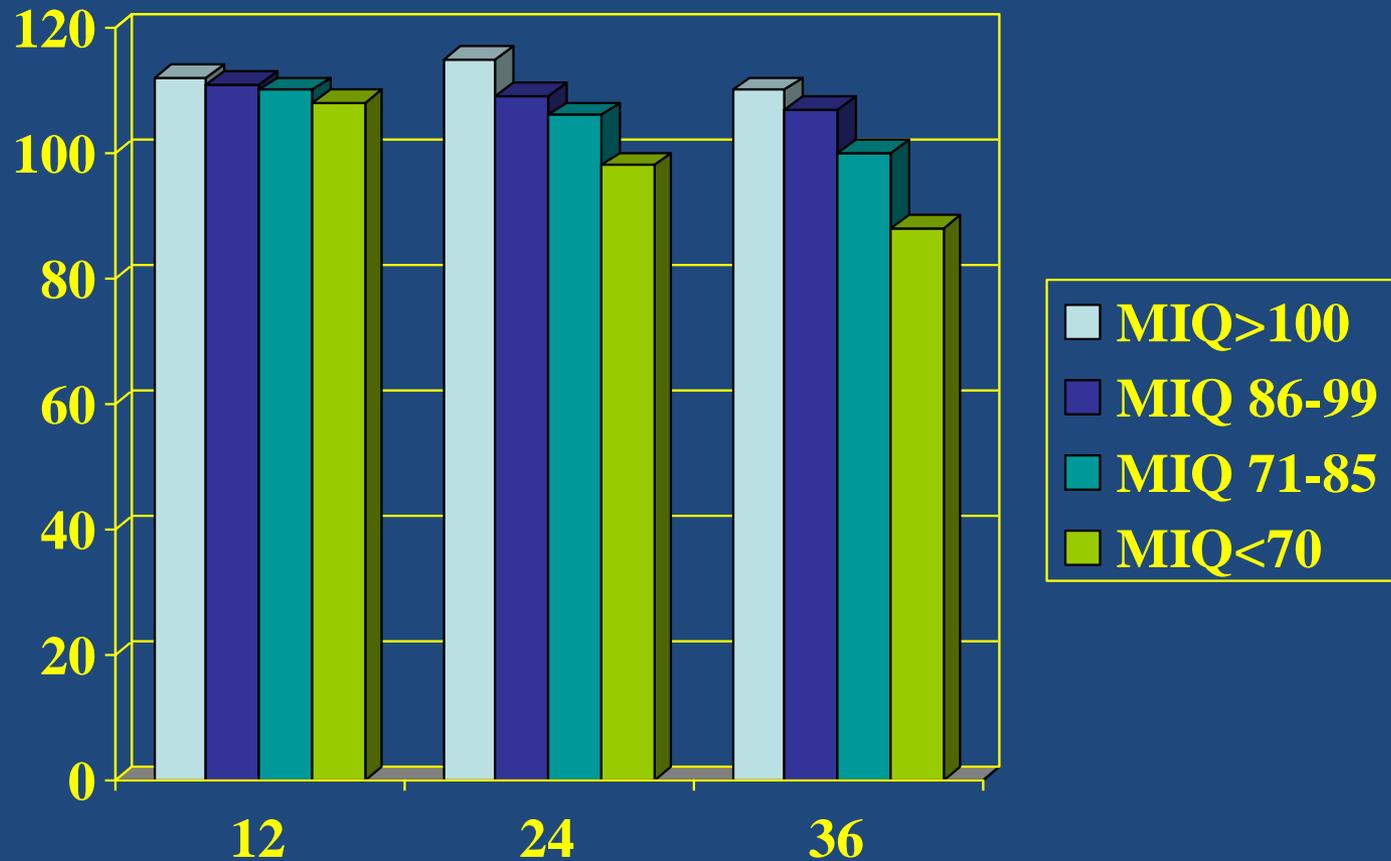


Percentage of children at age 3 years with cognitive impairment (IQ ≤ 85) and intellectual disability (IQ ≤ 70) in the IHDP

Child Cognitive Outcomes and Maternal(M) IQ: IHDP Controls



Child Cognitive Outcomes and Maternal IQ: Developmental Investment Group



VLBW and IHDP

	Intervention (N=98)	Control (N=182)
CP @3Y	10%	10%
SB IQ	88	80**
No CP-SB IQ	92	82***

Policy Impact of the IHDP

- Psychosocial intellectual disability can be decreased by comprehensive services
- At age 3, 87% of participants were in quality Head Start or preschool education in the community
- Long term impact on high school performance, risk behaviors, and outcomes was positive
- A strategy of parent engagement and supports for activity-based interventions promotes resilience.

Lessons from Epidemiology

- Risk ≠ Disability: multicausal pathways underlie the syndromes of cerebral palsy, intellectual disability and autism spectrum disorders
- Major etiologies include pre-, peri-, post-natal, multifactorial, and unknown events.
- We are entering an era of promise where we can analyze biomarkers, brain structure & function, neuroprotection, and developmental resiliency.
- However, unless we address the silos of medical, educational, and social information we will miss the possibilities of resilience.





Pathways Neurodevelopmental Ability CP Research and Implementation Model

1. Examine sequential outcomes after implementing translational science and community interventions so that disability is prevented, function optimized, and musculoskeletal integrity maximized
2. Our vision is to create systems of care that optimize life-course resilience, support families and empower neurodiversity
3. We will collaborate to inform population strategies that promote hope, widely disseminate best practices, and demonstrate sustainability and value.

Adaptive/Functional Assessments

The Warner Initial Developmental Evaluation of Adaptive and Functional Skills™

(WIDEA-FS™) • Version 12 • June 1, 2006

Michael E. Msall, Nancy Lyon, Larry Gray, Kathleen Mariano

»»» How often can your child do the following without help?

1 = Never 2 = Sometimes, infrequent 3 = Most of the time 4 = All the time

I. Self-Care: Feeding	
1. Easily drinks formula or breast milk	
2. Easily swallows baby food	
3. Chews solid food	
4. Finger feeds	
5. Eats using a spoon	
6. Drinks from cup without a lid	
7. Eats using a fork	
II. Self-Care: Dressing	
1. Holds arms up so you can put shirt on	
2. Removes socks	
3. Pulls pants down	
4. Pulls up a zipper once it is started	
5. Puts on t-shirt	
6. Removes all clothes	
III. Self-Care: Diaper Awareness	
1. Indicates a wet diaper	
2. Indicates a soiled diaper	
3. Voids into potty chair or toilet	
4. Sits on potty chair and has bowel movement	
Subtotal Self-Care Domain (max=68)	

IV. Mobility	
1. Rolls both ways	
2. Maintains sitting without support	
3. Crawls short distance	
4. Walks few feet with assistance (cruises)	
5. Scoots or moves in wheelchair 10 feet	
6. Walks 10 feet independently	
7. Crawls up stairs	
8. Gets on and off a chair	
9. Walks up stairs with hand held	
Subtotal Mobility Domain (max=36)	

V. Communication	
1. Understands words for people in immediate family (mommy, daddy) (R)	
2. Demonstrates 2 syllable babbling (E)	
3. Understands words for some common objects (R)	
4. Gestures a social greeting (wave, blow a kiss) (E)	
5. Carries out a 1 step oral request with gesture (pick up toy, cup) (R)	
6. Uses single words or signs to request or communicate (E)	
7. Carries out a 1 step oral request without gesture (R)	
8. Identifies one body part (R)	
9. Identifies three or more body parts (R)	
10. Points at pictures (R)	
11. Has at least 10 words or 10 signs (E)	
12. Combines words or signs to make needs known (E)	
13. Names pictures (E)	
Subtotal Communication Domain (max=52)	

VI. Social Cognition	
1. Plays "peek-a-boo", "patty cake", or "so big"	
2. Looks for object dropped out of sight	
3. Initiates social contacts with peers	
4. Takes turns rolling a ball	
5. Imitates another child	
6. Recognizes familiar song	
7. Starts mechanical toy or VCR/DVD/computer	
8. Can pretend play with doll or toy	
9. Turns pages in a book	
10. Points at pictures when you read a story	
11. Helps with simple household tasks	
Subtotal Social Cognition Domain (max=44)	

TOTAL WIDEA-FS SCORE

WIDEA-FS 4-18 MONTHS

