NEURODEVELOPMENTAL OUTCOMES IN HI: CHOP DATA

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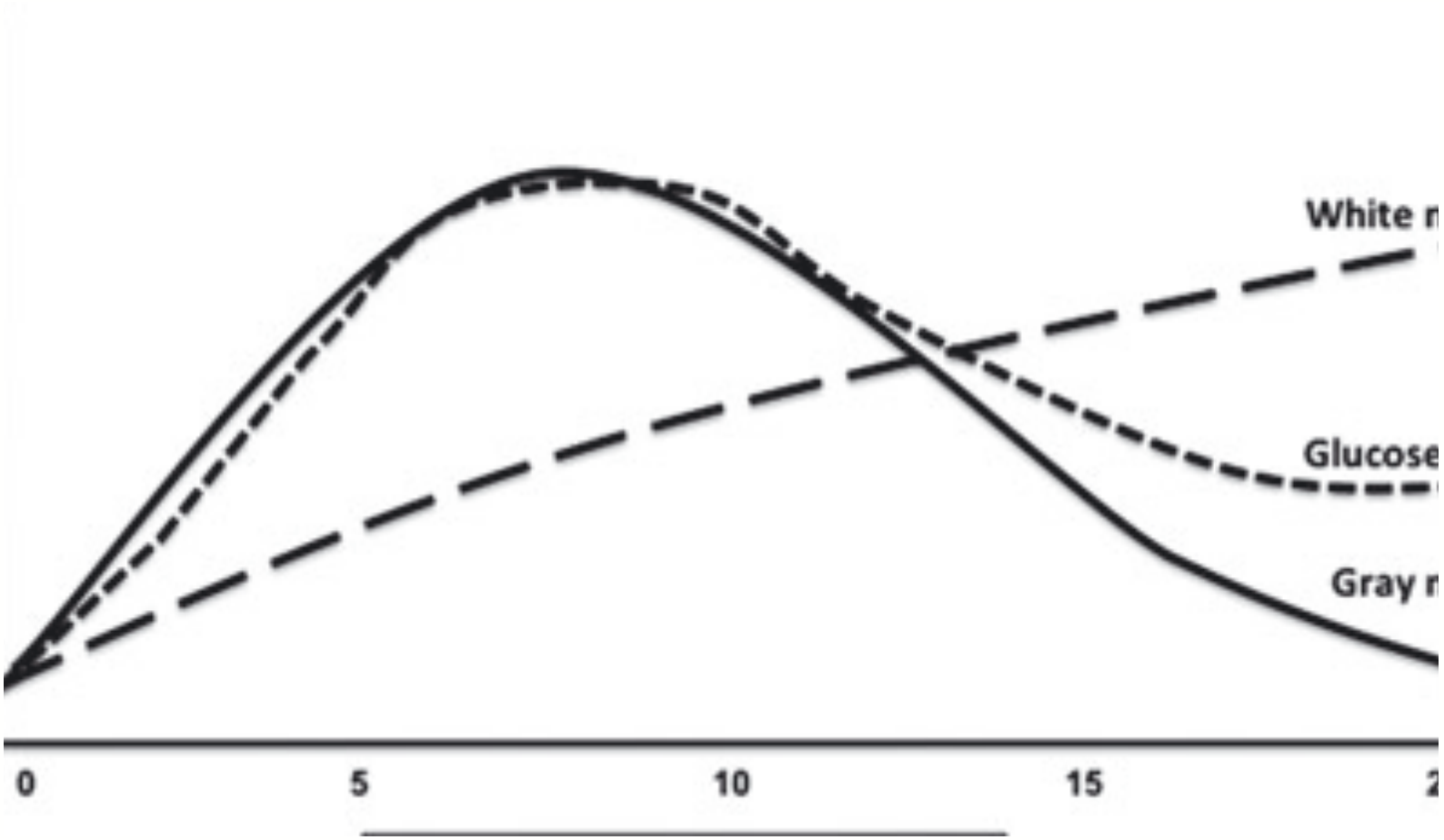
HYPOGLYCEMIA AND THE BRAIN

- Hypoglycemia can damage cells in the brain

- Children with HI are at risk of neurological complications
  - Developmental delays
  - Learning disabilities
  - Epilepsy

BRAIN DEVELOPMENT AND GLUCOSE NEEDS



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EFFECTS OF HYPOGLYCEMIA ON DEVELOPMENTAL OUTCOMES IN CHILDREN WITH CONGENITAL HYPERINSULINISM
CHOP 1980-2000

- 68 subjects
  - 35 who had pancreatectomy
  - 26 on medical treatment
  - 7 with transient HI, which they had outgrown
- History of Hypoglycemia Questionnaire
- Developmental Testing: Scales of Independent Behavior Revised

Steinkrauss, Lipman, Hendell, Gerdes, Thornton, Stanley, *J Pediatr Nurs* , 2005

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DEVELOPMENTAL OUTCOMES

	Average-Above Average (%)	Low-Low Average (%)	Very Low (%)
Overall	69	15	16
Surgical	68	9	23
Medical	73	23	4
Transient	57	14	29

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LONG-TERM OUTCOMES IN INDIVIDUALS WITH SURGICALLY-TREATED HYPERINSULINISM
CHOP 1960-2008

- 121 subjects who underwent pancreatectomy for HI
  - Prevalence of diabetes and neurological deficits
- Parent interview/questionnaire
- Developmental testing
  - Adaptive Behavior Assessment System (ABAS II)
  - Child Behavior Checklist (CBCL)

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Lord, Radcliffe, Gallagher, Adzick, Stanley, De León, *JCEM*, 2015

RESULTS OF DEVELOPMENTAL TESTS

Table 4. Neurobehavioral Measures

ABAS-II (n = 69)[a]	Mean ± SD	% < 1 SD	% < 2 SD
GAC score	96 ± 25	27.5[d]	18.8[e]
Conceptual composite score	98 ± 22	21.2	11.8[e]
Social composite score	100 ± 21	22.1	14.7[e]
Practical composite score	92 ± 25	30.9[e]	16.2[e]
CBCL (n = 62)[b]		% > 1 SD	% > 2 SD
TP score	49 ± 16	16.1	8.1[d]
Internalizing problems	49 ± 13	16.1	9.7[e]
Externalizing problems	47 ± 11	11.5	6.5[c]

Subjects scoring low on testing = 27%

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PARENT-REPORTED NEUROBEHAVIORAL PROBLEMS

Type	HI Patients	US Population
Psychiatric/behavioral	21%	13%
Speech delay	18%	8%
Learning disability	16%	8%
Seizures	13%	1%
Physical disability	11%	5%
ADHD	10%	7%
Autism	2%	0.5%
Any Problem	48%	

RISK FACTORS FOR NEURODEVELOPMENTAL ISSUES

- Factors not associated with developmental issues
  - Gender
  - Age at presentation
  - History of seizures at presentation
  - Age at surgery
  - Extent pancreatectomy
  - HI genetics

- Children with diffuse HI more likely to have lower scores on developmental testing than those with focal HI

- More research is needed

CONCLUSIONS

- Children with HI are at risk of developmental issues

- How to improve developmental outcomes
  - Early diagnosis and appropriate treatment with goal of maintaining normal blood glucoses
  - Developmental screening for all patients diagnosed with HI
  - Initiation of therapy if delays are identified

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NEUROPSYCHOLOGICAL ASSESSMENT AND EVALUATION

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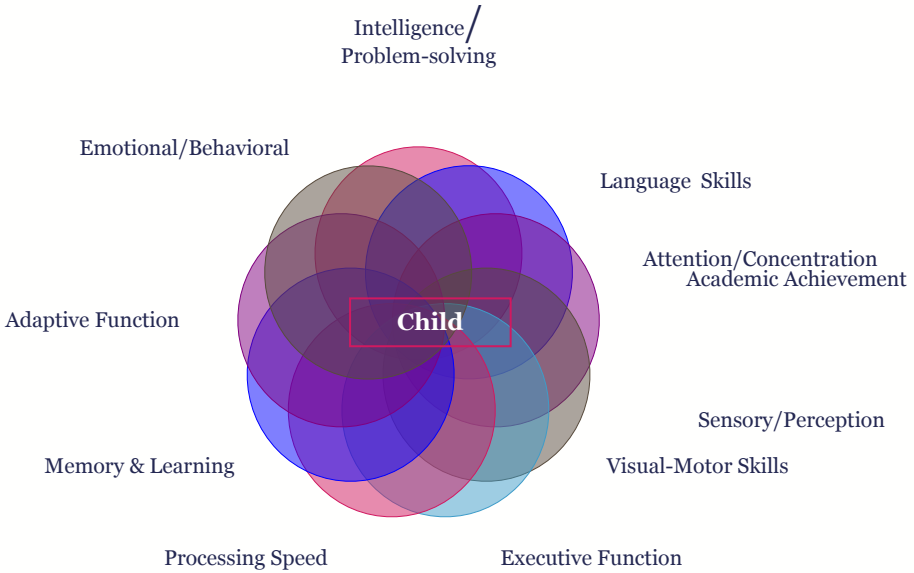
WHAT IS A NEUROPSYCHOLOGICAL EVALUATION?

- Comprehensive assessment using objective, standardized tests and procedures that compare performance of a child to a representative sample of same age peers
- Use brain-behavior relationships to explain functioning
- Typical neurocognitive domains assessed:
  - Intellectual abilities
  - Speech and language skills
  - Attentional regulation
  - Executive functions (planning, organization)
  - Memory and learning
  - Visual-perceptual/visual-spatial functioning
  - Visual-motor & fine motor skills
  - Academic achievement
  - Adaptive functioning
  - Behavioral and emotional status

WHAT ARE THE GOALS OF NEUROPSYCHOLOGICAL EVALUATION?

- Provide an explanation for difficulties a child might be experiencing
  - Writing difficulties: fine motor weaknesses, visual motor integration impairments, inattention, trouble generating ideas or organizing thoughts
- Clarify what is wrong (e.g., attention difficulties versus memory difficulties)
- Provide or confirm a diagnosis (e.g., learning disability)
- Identify cognitive strengths and weaknesses
- Document improvement or deterioration in conditions
- Provide recommendations to facilitate patient care and intervention planning

- Comprehensive
- Process focused
- Brain-behavior oriented



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COMPONENTS OF NEUROPSYCHOLOGICAL EVALUATION

- Record Review – medical & school records, past assessments
- Interview with caregivers – birth, developmental, family, school & psychosocial history
- Testing (4-7 hours; 1-2 sessions)
  - Behavioral observation
  - Examiner administered tasks
  - Computerized tasks
  - Parent and teacher questionnaires
- Feedback session, written report

WHAT HAPPENS NEXT?

- Neuropsychological report
  - Referral question
  - Relevant history
  - Review of behavioral observations and test results
  - Summary
    - Diagnostic impressions
    - Description of the individual's neurobehavioral/ neurocognitive profile
    - Strengths and weaknesses
    - Risk and protective factors
  - Recommendations

WHAT KIND OF RECOMMENDATIONS?

- Services at school
  - Individualized Education Program
    - Involves requesting a multidisciplinary evaluation from the school
    - Covered by Individuals with Disabilities Education Act (IDEA) 2004
  - 504 Plan
    - Covered by Section 504 of the Rehabilitation Act of 1973
  - School placement: classroom type, level of services, 1:1 aide
  - Special instruction in a particular subject
- Modification of school instruction
  - Extended time on assignments/tests
  - Reduction in homework volume, writing requirements
  - Organizational help

WHAT KIND OF RECOMMENDATIONS?

- Related Services- OT, PT, SL, hearing, vision services
- Therapy- school-based, private, parent training, social skills groups
- Psychiatric or neurological consult
- Resources- support groups, agencies, vocational rehab, specific reading materials

QUESTIONS?

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