Neurology and development in children with congenital hyperinsulinism

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Glucose and the brain

- The brain accounts for 20% of the body’s resting energy usage; more in children
- Glucose is essential for brain function and growth
- Low glucose levels can reduce brain function
- If glucose levels stay too low for too long it can injure the brain
Anatomy of the brain
Anatomy of the brain
Brain function depends on electrical and chemical messages passed through nerve cells and their connections. Glucose is a fuel for creating these messages.
The brain in children: early beginning
The brain as the infant develops
Changes in the brain through adolescence
Changes in body and function in childhood
Specific developmental domains and trajectories

Vision

Fine motor

Social rules

Infant  Childhood  Teenager  Adult
Severe hypoglycaemic brain injury
Consequences of severe hypoglycaemic brain injury

• Cerebral palsy
• Epilepsy
• Visual impairment
• Learning disability
Cerebral palsy
Child with Lennox-Gastaut Syndrome pattern

10 year old, early prolonged hypoglycaemia, evolved four limb disability with learning difficulties, ongoing admissions with polymorphic seizures
Vision and visual pathways
Milder effects on higher neurocognitive functions

- Fine motor control
- Higher visual perception
- Attention, concentration, memory
- Reading, spelling, arithmetic
Fine motor functions
Assessing visual effects
Assessing milder higher neurocognitive functions
Academic skills
Higher cognitive abilities

Memory

Concentration

Planning
Assessments

• Vision assessment, sensory impairment teacher
• Educational psychology and/or neuropsychology assessment
• Occupational therapy
• Physiotherapy
• Speech and language therapy
Developmental outcomes in hyperinsulinism

• 2017 study from Manchester
• 12% had low scores on a development questionnaire
• More detailed tests show more affected
• Males tended to have lower scores
• Late presenting CHI tended to have lower scores
• Diazoxide-unresponsiveness and genetic mutations had lower scores
Fig. 1 VABS-II scores have been expressed as SDS in patients with Early-CHI and Late-CHI for individual and adaptive behaviour composite (Total) domains. The reference line at 0 SDS represents the population mean with values < -2.0 representing significant deviation.
Summary

- A tour of typical brain anatomy and function
- Typical development in childhood
- How development and problems are assessed
- Emerging developmental findings in congenital hyperinsulinism