Neurodevelopmental Outcomes In Children with Early and Late Presenting Congenital Hyperinsulinism

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Introduction

- Adverse neurodevelopmental outcomes have been recognised in children with hypoglycaemia due to early and late presenting Congenital Hyperinsulinism (CHI). However, specific phenotypes of adverse neurodevelopment have not been clearly described. We have used the Vineland Adaptive Behaviour Scales II (VABS-II) questionnaire to identify patterns of adverse neurodevelopment and the Social Communication Questionnaire (SCQ) as a screening instrument for Autistic Spectrum Disorders.
- VABS-II is a parent reported standardised tool used to assess adaptive behaviour in 5 domains: motor, communication, daily living skills, socialisation and maladaptive behaviour.
- The SCQ is a carer reported assessment tool used to screen for neurodevelopmental difficulties associated with an autistic spectrum disorder (ASD). SCQ provides a cut-off score that can be used to indicate the likelihood that an individual has an ASD, prompting early intervention.

Figure 1: VABS-II scores expressed as SDS plotted against age at presentation of CHI.

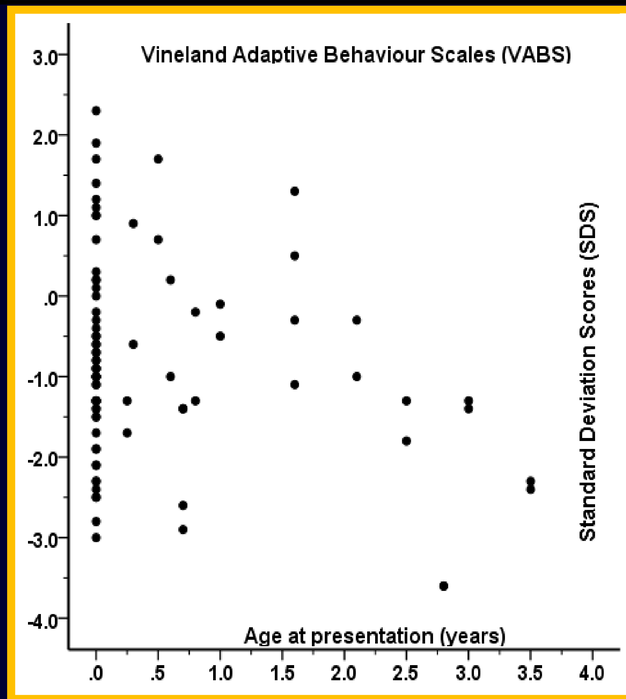
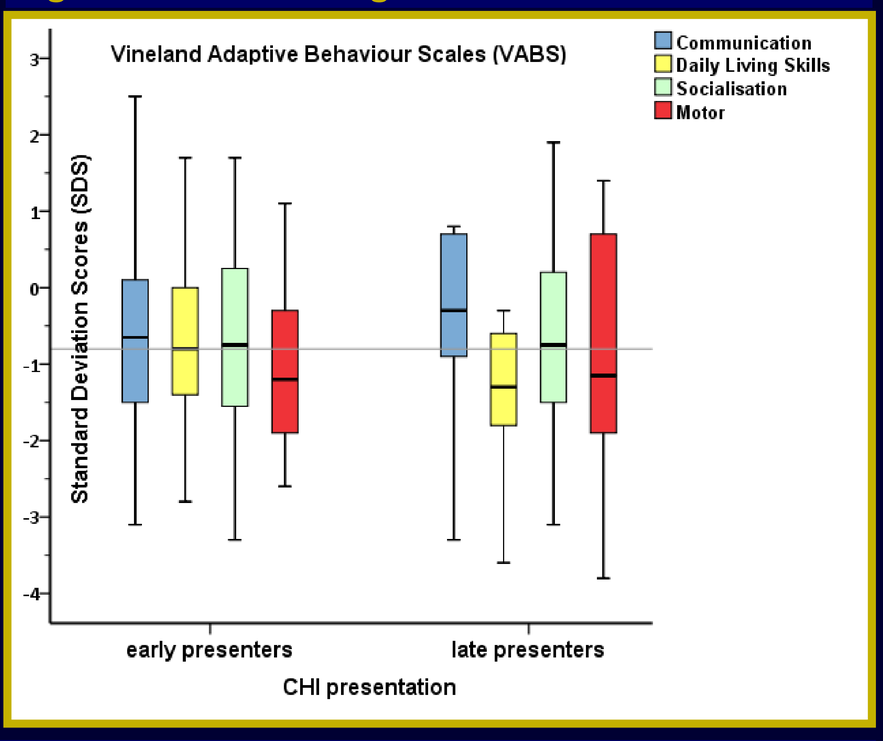


Figure 2 – VABS-II domains in early and late CHI: higher SDS indicating better outcome



Methods

- A cohort of 42 children (29 males, 69%) with CHI treated either medically surgically, was identified at age > 1.5 years.
- Those presenting with CHI before age 1 month were early CHI, while those presenting after 1 month were late CHI.
- Total VABS-II and sub-domain scores except behaviour were converted to standard deviation scores (SDS) using normative data.
- SCQ outcomes were reported as raw scores; children with scores > 15 were referred for formal testing for autism spectrum disorders (ASD).

Figure 4 – SCQ raw scores in early and late CHI

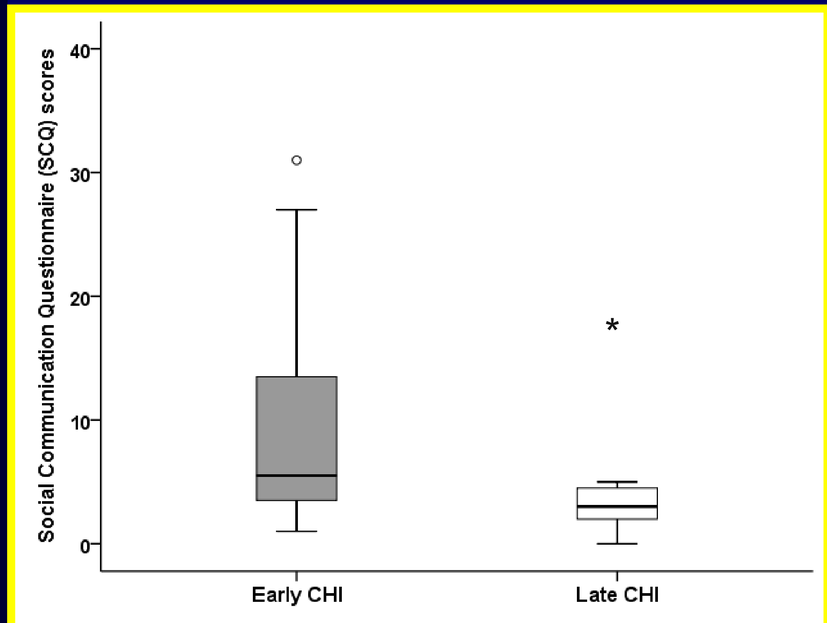
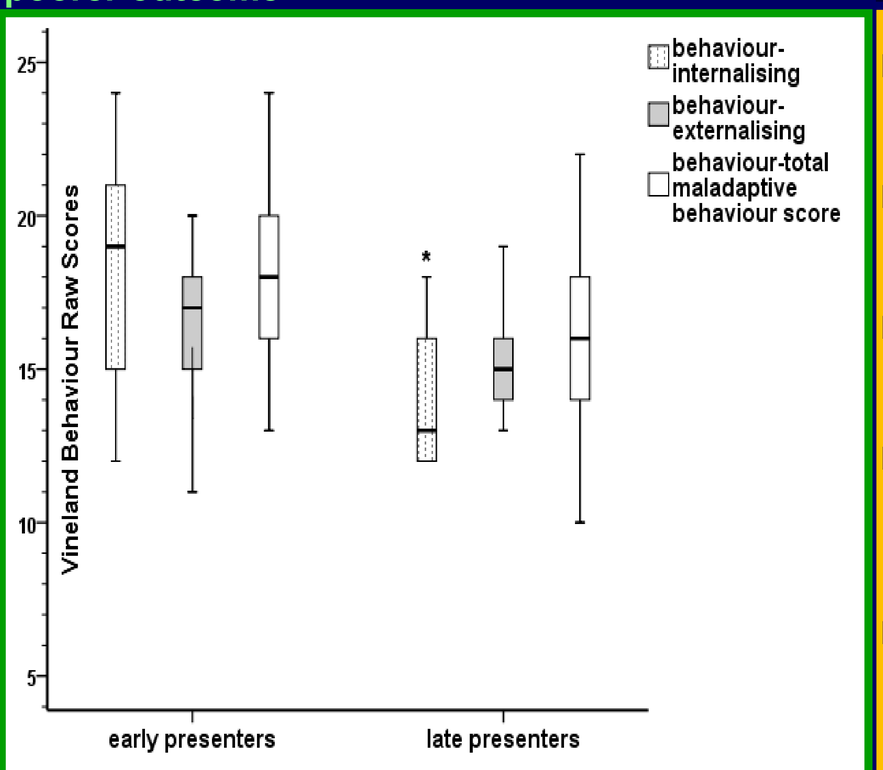


Figure 3– VABS-II maladaptive behaviour domain in early and late CHI: higher raw scores indicating poorer outcome



Results

- In our CHI cohort, 27(64%) children presented early, while 15 (36%) presented later [median (range) age 1.0 (0.3; 3.5) years].
- VABS-II-SDS was low in the whole cohort [-1.0 (-3.6;+2.3)] and was negatively correlated with age at presentation [R=-0.3, p=0.05] [Figure 1].
- The Daily Living Skills (DLS) domain was significantly lower in late CHI, despite being adjusted for gender CHI [Figure 2, p=0.04].
- In contrast, early CHI showed a higher behavioural score than late CHI [18 (13;24) v 16 (10;22), p=0.1], particularly for internalising behaviours [p=0.02], suggesting maladaptive behaviour in early presenters [Figure 3].
- SCQ scores were also higher in early CHI than late CHI [5.5 (1;31) v 3.0 (0;18), p=0.04], suggesting the tendency to an autistic spectrum disorder (ASD) with early presentation. Of those referred for formal testing for ASD, autism was diagnosed in 1 child, each from early and late CHI.

Conclusion

- **VABS-II psychometric testing shows differential neurodevelopmental phenotypes in early and late CHI, suggesting that the pattern of hypoglycaemic neuronal injury is dependent on the age at presentation.**