The role of plasma C Peptide concentration in the diagnosis of Congenital Hyperinsulinism (CHI)

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BACKGROUND
• The hallmark of CHI is the demonstration of detectable plasma insulin during hypoglycaemia
• Plasma insulin can be undetectable in a significant proportion of patients with CHI
• Plasma samples for insulin requires rapid and careful handling for reliable results
• There is little published data on the value of plasma C Peptide in the diagnosis of CHI

OBJECTIVE
To assess the usefulness of plasma c peptide in the diagnosis of CHI

METHOD
• All completed hypoglycaemia (laboratory glucose <2.6 mmol/l) screening tests undertaken at a tertiary referral centre over a 4 year period were assessed retrospectively
• The diagnosis of CHI was made on a combination of glucose requirement >8mg/kg/min, detectable insulin during hypoglycaemia, suppressed ketones and a glycaemic response to glucagon
• The plasma C peptide concentration during hypoglycaemia in patients with CHI was compared with that of the patients with other diagnoses (including peroxisomal disorders, respiratory chain disorders, growth hormone deficiency and ketotic hypoglycaemia).

RESULTS
• 60 results were available from 41 patients.
• Median age was 16 months (1day – 20.5 years). Diagnoses included 23 CHI, 1 growth hormone deficiency, 1 peroxisomal disorder, 1 respiratory chain disorder and 15 ketotic hypoglycaemia.
• The concentration of plasma C peptide in patients with CHI was significantly higher than that for patients with a diagnosis other than CHI as shown in Fig 1(p<0.001).
• The positive predictive value for a diagnosis of CHI with C peptide concentration of 350 pmol/l is 96.7%.
• The negative predictive value is 80%.

CONCLUSION
A plasma C peptide concentration of > 350 pmol/l during hypoglycaemia could be used to differentiate CHI from other causes of hypoglycaemia especially when plasma insulin level is low or could not obtained.

REFERENCE