

The University of Manchester



Towards more tolerable therapy for CHI

Indi Banerjee

Royal Manchester Children's Hospital

Treatment options in CHI

- Diazoxide, octreotide, lanreotide
- Sirolimus

• Therapies in trial

• ? Supplementary therapy

Fish Oil - PolyUnsaturated Fatty Acids (PUFA)

- GISSI myocardial infarction trial PUFA reduces abnormal heart rhythms after heart attack GISSI investigators, Lancet 1999
- GISSI Heart Failure PUFA restores autonomic variability La Rovere et al, Heart Rhythm 2013
- GISSI Heart Failure PUFA reduces life threatening ventricular arrythmis Finzi AA, Am Heart J 2011
- Reduction in all-cause mortality after heart attack Green SJ et al, Am J Cardiology 2016
- Lowers mortality in adults with high risk of cardiovascular problems Guasch-Ferre M et al, Am J Clin Nutr 2015

Trial of PolyUnsaturated Fatty Acids (PUFA) in children treated with Diazoxide

MaxEPA (liquid) 3 mL/day [EPA 459 mg, DHA 310 mg] (n=13)



Modest effect in improving Sugar profiles (subcutaneous glucose)



Significant response in some children, e.g. in a child with a metabolic cause for Hyperinsulinism (HADH mutation)



Reduced risk of low blood sugars with PUFA treatment

Mechanism of better sugar levels?

 Calcium channels that cause insulin secretion in beta cells may become more resistant – may explain less hypoglycaemia

• Better beta cell membrane stability?

• Other effects?

PUFA effects in glucose metabolism







KIDS Omega 3 Syrup

 A super tasty syrup which supports healthy growth and development EPA 460 mg in 5 ml

ESKIMO-3 little cubs

Rids formula Omega-3 Fish Oil with Vitamin D & E

> range avour

od Suppleme

EPA 260 mg in 5 ml

Progressive dose reduction in diazoxide: 12 mg/kg/d to 1 mg/kg/d

Post trial PUFA use

- 9 patients with and without genetic mutations
- 4 children stopped medication
 - 2 children with no CHI mutations
 - 1 child with PHOX2B mutation
 - 1 child with homozygous ABCC8 mutation previously treated with lanreotide
- Others reducing diazoxide dosage



Reducing diazoxide dose to reduce excess body hair

Need to treat hypoglycaemia, while recognising Transient forms of CHI





CHI - Persistent or Transient



Understanding the basis of complications in CHI



Children's Hospital

University of

Manchester

Parents Accommodation

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A dl

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