



# British Care Guidelines

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# Standardised practices in the networked management of congenital hyperinsulinism: a UK national collaborative consensus

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Helen McDevitt<sup>1</sup>, Zainab Mohamed<sup>15</sup>, Kate Morgan<sup>2</sup>,  
Jacqueline Nicholson<sup>16</sup>, Ana Nikiforovski<sup>3</sup>, Elaine O'Shea<sup>3</sup>,  
Pratik Shah<sup>17</sup>, Kirsty Wilson<sup>1</sup>, Chris Worth<sup>3</sup>, Sarah Worthington<sup>3</sup>  
and Indraneel Banerjee<sup>3</sup>



# Aims



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**Consistency in High-Quality Care**

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**Efficient Diagnosis and Treatment**

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**Equitable Access**

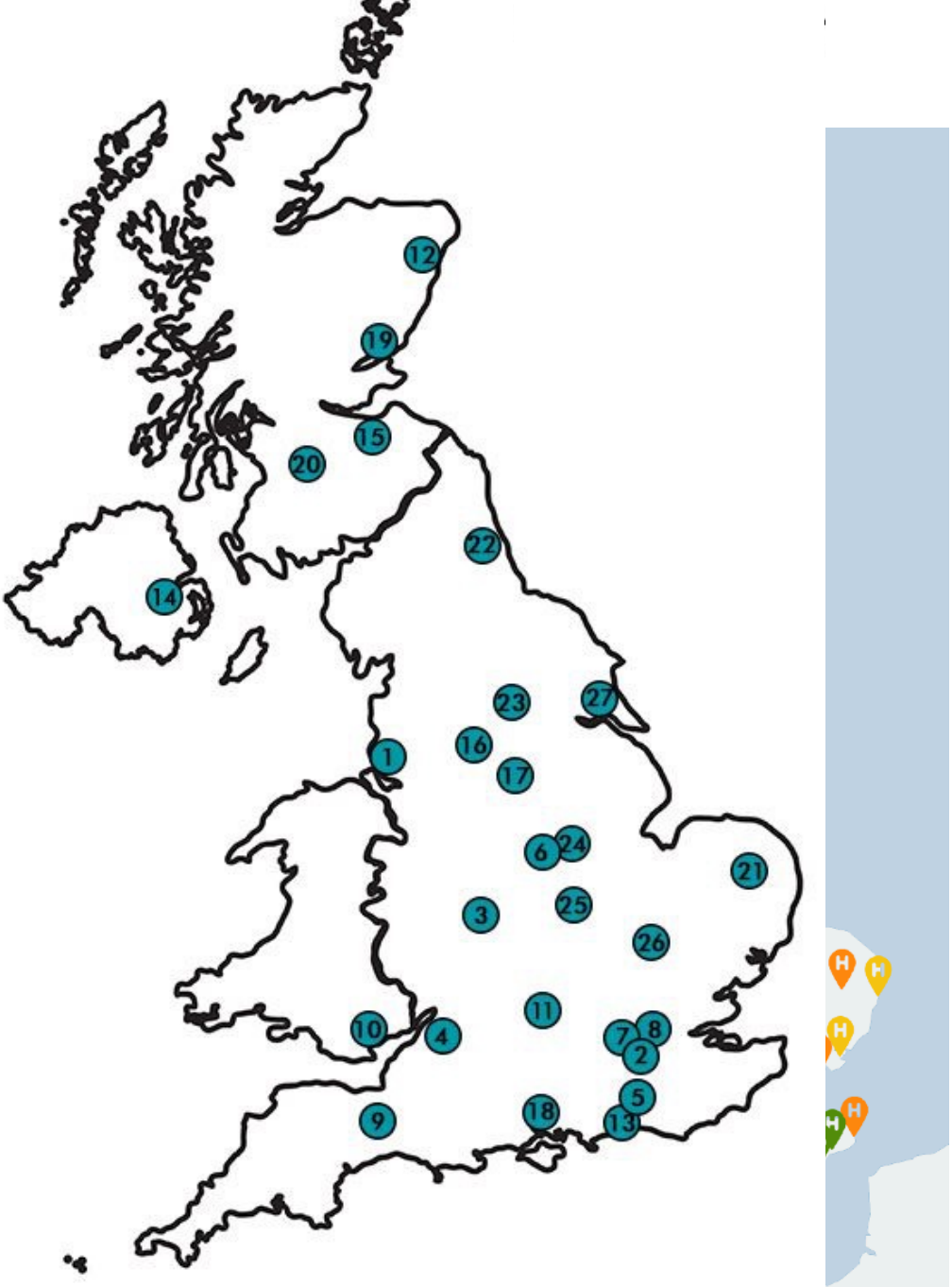
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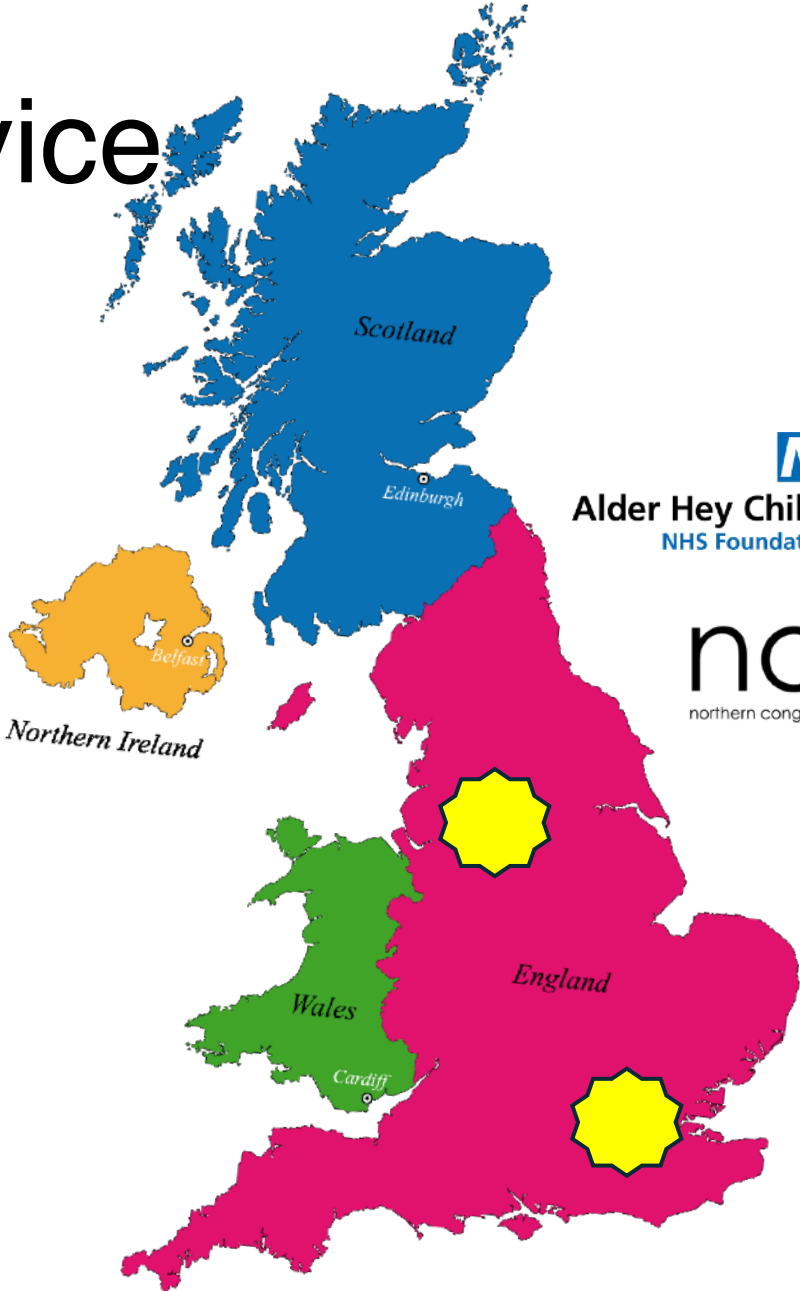
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**Enhanced Collaboration**

# National Health Service



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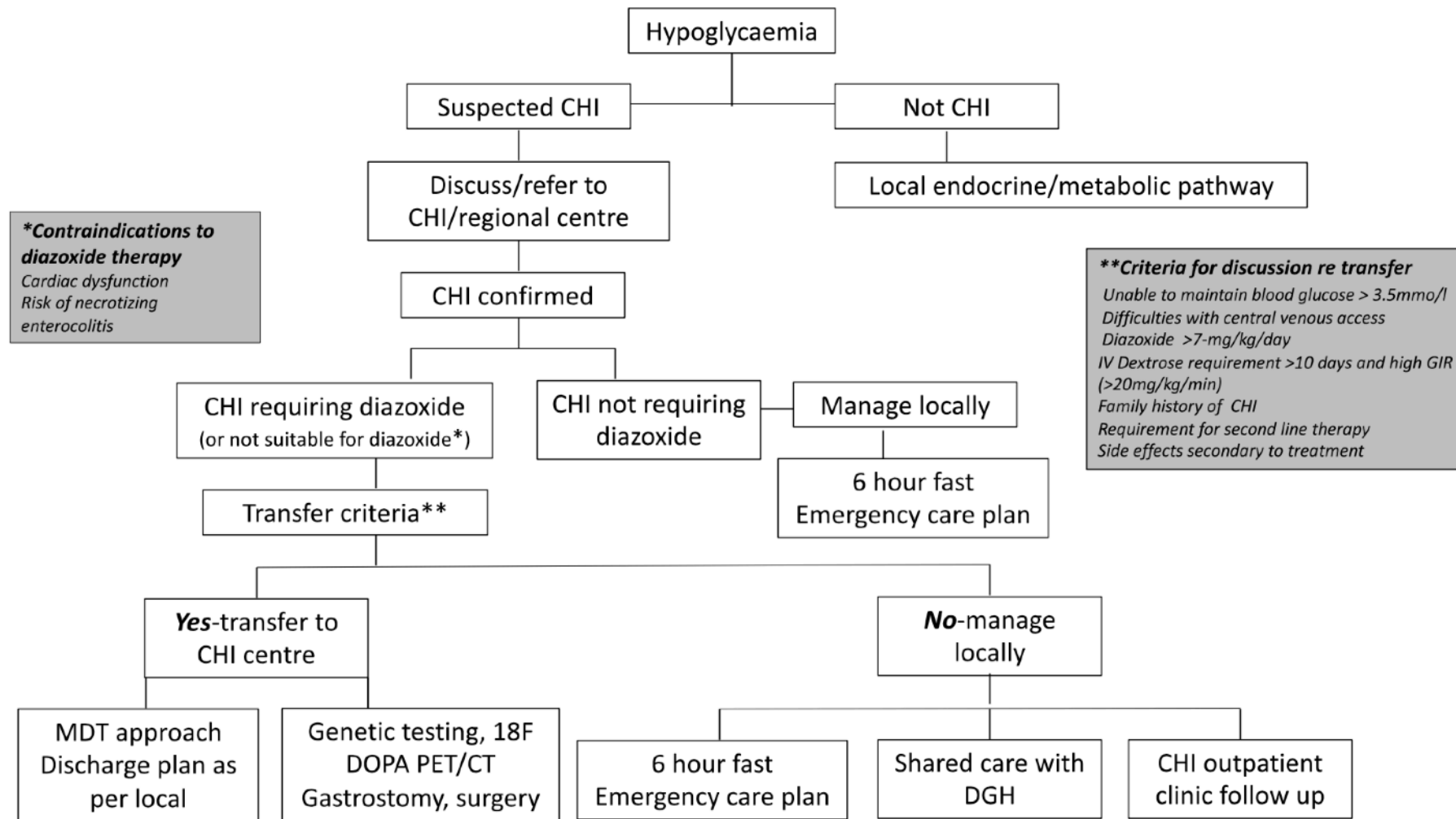
**NHS**  
Alder Hey Children's  
NHS Foundation Trust



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northern congenital hyperinsulinism service



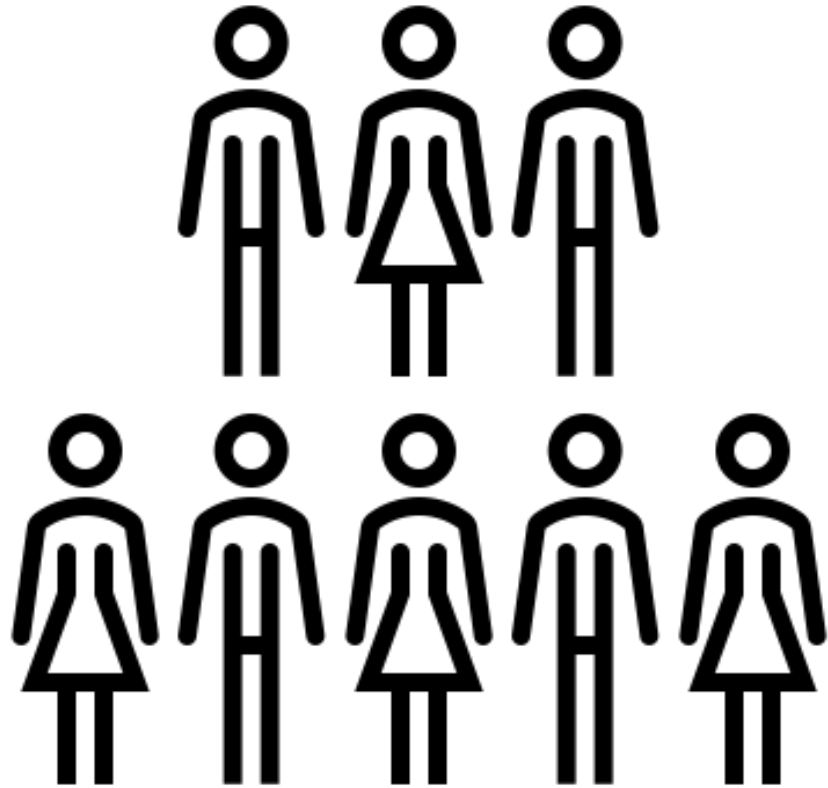
**NHS**  
Great Ormond Street  
Hospital for Children  
NHS Foundation Trust



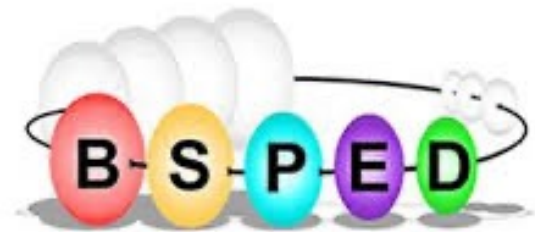
**FIGURE 1**

Referral pathways and criteria for congenital hyperinsulinism diagnosis and treatment in a networked model of care. CHI, congenital hyperinsulinism; DGH, district general hospital.

# Consensus Group



- CHI Special Interest Group
  - Subcommittee of the British Society for Paediatric Endocrinology and Diabetes (BSPED)
  - Healthcare professionals with extensive experience in CHI across the UK
  - Patient representatives from the Children's Hyperinsulinism Charity (CHC)
- Virtual meetings over 18 months (ending March 2023)



# Presentation and Diagnosis

- Threshold for investigation of hypoglycaemia of 3.0mmol/L (54 mg/dL)
- Severe and/or recurrent hypoglycaemia
- 2 or more episodes of glucose <3.0mmol/L with glucose infusion rate of >8mg/kg/min
- Suppressed ketones and fatty acids





# Acute management



Aim for glucose  $> 3.5\text{mmol/L}$  (63.0 mg/dL)



Dextrose boluses



High concentration glucose infusion  
(15-20%) via central venous catheter



Glucagon infusion

# Criteria for Potential Transfer to CHI Centre

Unable to maintain BG >3.5mmol/L

Difficulties with central venous access

Diazoxide >7mg/kg/day

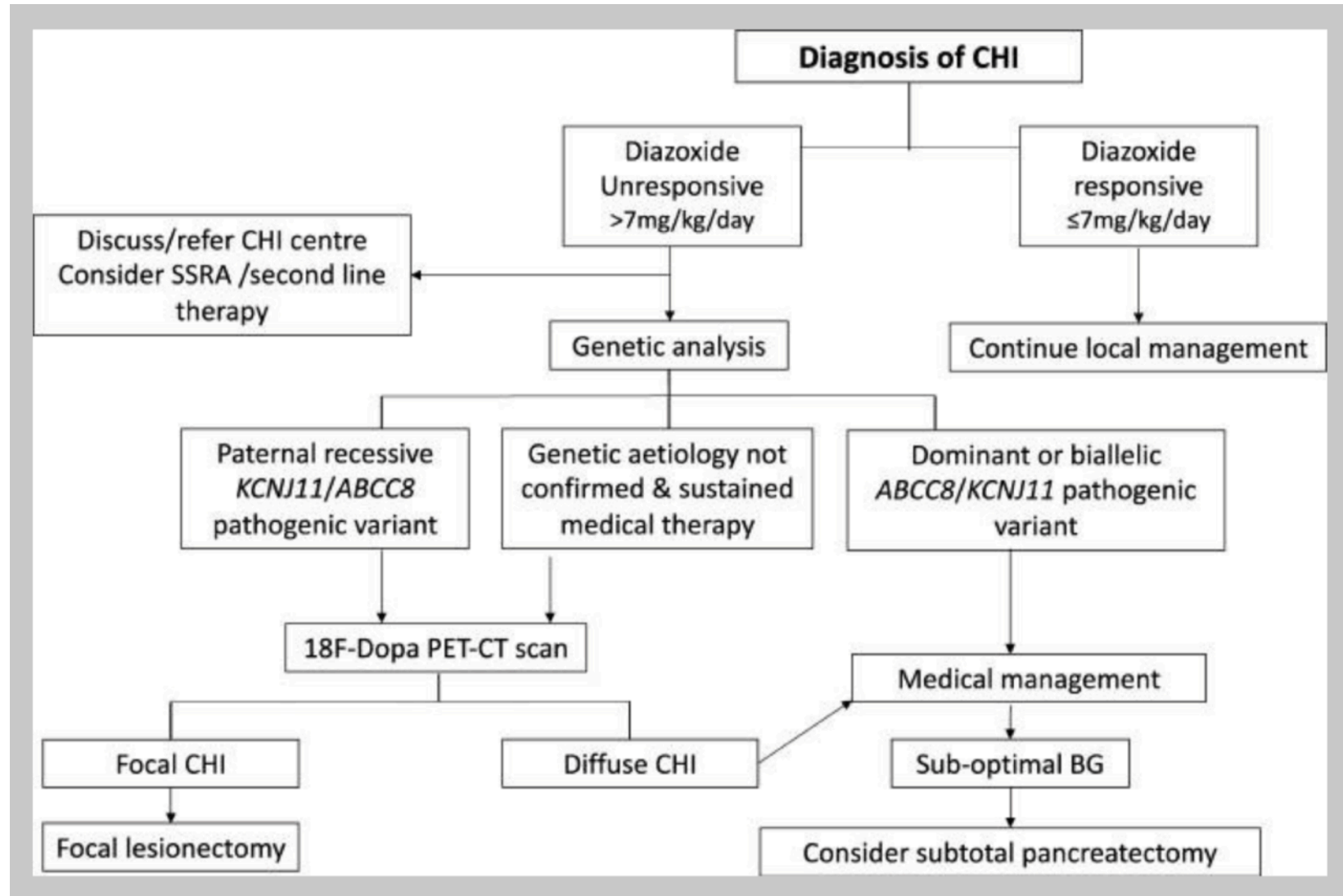
IV dextrose requirement >10 days and high GIR (>20mg/kg/min)

Family history of CHI

Requirement for second line therapy

Side effects secondary to treatment

# Treatment



Clinical pathway of diagnosis and management for patients with congenital hyperinsulinism (CHI). Clinical decisions hinge on criteria such as diazoxide responsiveness, genetic investigations, 18 Fluoro Dopa PET-CT/MR imaging and response to medical therapy.

# Feeds

- Refer to dietitian if requiring additional oral carbohydrate to prevent hypoglycaemia
- Monitor protein/energy ratio to achieve adequate growth
- Proportion of patients with feeding problems
  - Requires early recognition and MDT input including SALT and dietetics



# Blood glucose monitoring

- Frequency of monitoring
- Continuous glucose monitoring (CGM) alongside point of care tests in hospital
- CGM at home for those in need
  - Useful for pattern recognition to motivate behavioural changes rather than for acute detection of hypoglycaemia



# Long term Management

- Outpatient review 3-6 monthly
- More frequent virtual reviews often required to optimise therapy
- Minimal treatment -> consider treatment withdrawal, age-appropriate safety fast, satisfactory profile on home glucose monitoring
- If resolved, consider infrequent follow up

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# Summary

- Guidelines created with aim of standardising practice across UK
- Aim for efficient diagnosis and treatment
- Networked care in the UK
- Acute management
- Criteria to consider transfer to CHI centre
- Long term management of feeds, blood glucose monitoring etc



# References

- Shaikh MG *et al.* Standardised practices in the networked management of congenital hyperinsulinism: a UK national collaborative consensus. *Front Endocrinol (Lausanne)*. 2023 Oct 30;14:1231043. doi: 10.3389/fendo.2023.1231043. PMID: 38027197; PMCID: PMC10646160.
- <https://www.norchi.nhs.uk>
- <https://www.gosh.nhs.uk/conditions-and-treatments/conditions-we-treat/hyperinsulinism/>