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Case Study: Nephrogenic Diabetes Insipidus (NDI) management simplified by the use of Renastep™

Highly Specialised Paediatric Dietitian, UK

Patient Details & Medical History

Age:
3 years

Gender:

Diagnosis:
Nephrogenic Diabetes Insipidus. Mutation in AVPR2.

Medical history:

Premature 27/40, birth weight 880g, polyuria, hypernatraemia.
Diagnosis of NDI suspected due to family history. Confirmed genetically on day 20.

Dietetic history:

- Initially received parenteral nutrition. Weaned off by day 35. Expressed breast milk (EBM) or low renal solute load (RSL) feed (dilute infant formula) given orally or via nasogastric (NG) tube.
- Required continuous NG feeding due to vomiting. Gastrostomy inserted at 23 weeks corrected gestational age.

At 3 years of age (no longer correcting for prematurity):

Weight: below -5SD (<0.4th centile)[1] slow gain - tracking.

Length: below -5SD (<0.4th centile)[1] static.

Biochemistry at current presentation:

Component	Results	Normal range	Comment
Serum			
Sodium	**153 mmol/L**	133 - 146 mmol/L	Hypernatraemia (dehydrated)
Potassium	3.9 mmol/L	3.5 - 5.5 mmol/L	
Total CO_2	25 mmol/L	20 - 30 mmol/L	Not acidotic
Urea	4.5 mmol/L	2.5 - 6.0 mmol/L	
Creatinine	16 mmol/L	23 - 37 umol/L	Low muscle mass. Normal renal function
Calcium	2.57 mmol/L	2.22 - 2.51 mmol/L	Possibly reflecting dehydration
Phosphate	1.69 mmol/L	1.2 - 1.8 mmol/L	
Osmolality	**322 m0smol/kg H_2O**	282 - 300 m0smol/kg H_2O	High
Urine			
Osmolality	**163 m0smol/kg H_2O**	Interpreted along with paired serum value	Inappropriately low vs serum value

This biochemistry suggests hypernatraemic dehydration. The renal function is otherwise normal (with no increase in serum creatinine or acidosis). The paired serum and urine osmolality values show that the child is unable to fully concentrate their urine to correct this dehydration. A child with no underlying health condition would increase their urine osmolality if dehydrated to retain more water which would normalise serum osmolality and serum sodium. This suggests the diagnosis of diabetes insipidus which genetically has been confirmed to be of renal rather than central origin (i.e. nephrogenic diabetes insipidus).

Patient Details & Medical History

Current medication:

Non-steroidal anti-inflammatory (for NDI)	Suppositories (to manage constipation)
Thiazide diuretic (for NDI)	IV fluids (started by central line to improve hydration)
Proton pump inhibitor (gastro protection)	

Current presentation:

- Remained on continuous gastrostomy feeds over 20 hours of diluted infant formula (¾ normal strength).
- Nil orally and recurrent vomiting with increased feed rate / concentration / energy density.

Dietetic Assessment

Overall aim/goal:

- To lower renal solute load of feed to reduce fluid requirements and to possibly reduce the need for IV fluids.
- To reduce duration of continuous gastrostomy feed (by increasing feed rate/concentration).
- To encourage oral intake and progression of feeding development.
- To improve nutritional intake to achieve better growth and weight gain.

Current management:

Weight: 7.2 kg (<0.4th centile. Z score below -5SD)[1].

Theoretical requirements for height age (10 months)**:** Energy >72 kcal/kg body weight (SACN Estimated Average Requirements EAR)[2]; protein 1.6g/kg body weight (Reference Nutrient Intake RNI)[3].

Feed: ¾ strength powdered infant formula - 1660 ml fed via gastrostomy at 83 ml/hr over 20 hours.

Fluids: 300 ml/kg (feed 230 ml/kg; IV fluids 70 ml/kg).

Nutrition: 100 ml ≈ 48 kcal, 1g protein.
1660 ml ≈ 799 kcal, 15.9g protein, 11.5 mmol Na, 22 mmol K.
Per kg ≈ 111 kcal, 2.2g protein, 1.6 mmol Na, 3.1 mmol K.

RSL: 18 m0smol/kg (aiming for ≤15 m0smol/kg).

Management plan:

Plan: To trial change of feed from ¾ strength powdered infant formula to Renastep (made to a 25% concentration with water - see the feed details below).
Lower renal solute load; easier to administer; potential for future oral intake.

Feed: Renastep (at a 25% concentration) - 1660 ml fed via gastrostomy at 83 ml/hr over 20 hours.

Fluid aim (enteral): 230 ml/kg.

Nutrition: 100 ml ≈ 50 kcal, 1g protein.
1660 ml ≈ 830 kcal, 16.6g protein, 14.9 mmol Na, 3.7 mmol K.
Per kg ≈ 115 kcal, 2.3g protein, 2.1 mmol Na, 0.5 mmol K.

RSL: 14.4 m0smol/kg.

Final aim: Mix 1 bottle of Renastep (125 ml) with 375 ml water to make 500 ml of 25% Renastep (Prepare x 4 per day).
Total feed 1660 ml (415 ml Renastep in total at a 25% concentration).
Gradual change over 1 week. Replacement of diluted infant formula with Renastep at a 25% concentration.

Results

Monitoring/review:

✓ **Tolerance:** Successfully changed over onto Renastep over 1 week – standard infant formula was stopped.

✓ **Checking serum electrolytes daily:** Serum sodium and osmolality returned to normal range.

Gradual ↓ in serum K to 3.3 mmol/l.

→ Started potassium sparing diuretic.

→ Replaced some feed with standard infant formula to provide additional potassium intake after 5 days.

Final feed:

Recipe: Family given simpler recipe for 24 hours: 2 bottles Renastep (250 ml in total), 100g infant formula powder made up to 2 L with water

Feed: 1660 ml fed via gastrostomy at 100 ml per hour over 16-17 hours.

Fluids: 230 ml/kg gastrostomy feeds.

Nutrition: 100 ml ≈ 49 kcal, 1g protein.
1660 ml ≈ 113 kcal/kg, 2.3g protein/kg.
100 ml ≈ 0.8 mmol Na, 0.8 mmol K.
1.8 mmol Na/kg, 1.8 mmol K/kg.

RSL: 16.3 m0smol/kg H_2O.

✓ Stable biochemistry (serum osmolality, sodium and potassium in normal range).

✓ Feeds given over 16 hours (no feeds between 9am and 5pm).

✓ Drinking water well and ↓IV fluids.

✓ Started to take solids ✓ No vomiting ✓ No constipation ✓ More settled

✓ Reported improved physical/psychological development as free from pump during the day and ↓thirst.

Weight and height: Increased to -4SD.

Discussion

The renal solute load of most ready to feed paediatric enteral formulae may be too high for underweight young children with NDI and this can pose a challenge.

Strengths, limitations or barriers:

- Renastep is low in potassium, sodium, phosphorus, calcium and vitamin A compared to standard paediatric enteral feeds. The main use of Renastep is for the dietary management of chronic kidney disease, so regular careful monitoring of overall nutritional intake is required to ensure requirements are met if used for other renal conditions.
- The ready to feed format of Renastep may simplify ongoing feed preparation and reduce the risk of preparation errors which can occur when reconstituting powdered feeds.

Conclusions

The low electrolyte content of Renastep may be useful when trying to reduce the RSL to aid progression of enteral feeds for young children with NDI.

A child with NDI on a feed providing a RSL >18 m0smol/kg who is not thriving, may struggle to have an appetite for food as they are so thirsty. Reducing the RSL while maintaining energy intake in this case, resulted in an improvement in oral solid and fluid intake.

Important Points

- The potassium content of Renastep is very low so careful monitoring of serum biochemistry is required.
- For future cases, a slower transition and titration of standard infant formula and Renastep would alert to decreasing serum potassium levels.

Reference:

1. RCPCH. 2012. UK-WHO growth charts - 2-18 years. [online]. Available from: https://www.rcpch.ac.uk/resources/uk-who-growth-charts-2-18-years
2. Scientific Advisory Committee on Nutrition. Dietary Reference Values for Energy. TSO, London, 2011.
3. Dietary Reference Values for Food Energy and Nutrients for the United Kingdom. Report on Health and Social Subjects No. 41. Department of Health, London, HMSO, 1991.

This information is intended for use by Healthcare Professionals only.

Renastep is a Food for Special Medical Purposes. Must only be used under strict medical supervision with regular monitoring of nutritional status and electrolyte levels. Renastep is a ready to use feed for the dietary management of kidney disease from 3 years of age onwards. For enteral use only. Not suitable as a sole source of nutrition.

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