Early enteral support in children with cystinosis: impact on growth.

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Introduction

- **Cystinosis** is a hereditary dysfunction of the renal tubules characterized by the presence of carbohydrates and amino acids in the urine, excessive urination, and low blood levels of potassium ions and phosphates.
- It is caused by the **accumulation** of cystine crystals in tissue.
- The renal Fanconi syndrome usually manifests by 4-6 months of age with polyuria,polydipsia, failure to thrive, vomiting, constipation, dehydration, rickets, in association with biochemical evidence of proximal tubular dysfunction.

Nutritional care

- Non- specific , symptomatic treatment of the renal Fanconi syndrome includes providing appropriate nutrition and substituting renal losses: these are crucial to allow **satisfactory growth**.

- Most young children with cystinosis develop severe progressive **failure to thrive**, unless they receive high caloric diets.

- **Early enteral support** should be considered in all children who have poor appetite or frequent vomiting.

Nephropatic cystinosis: an international consensus document. Nephrol Dial Transplant (2014)

Dietary reccommendations

Replacing the excessive urinary losses of amino acids, glucose, phosphate, calcium, magnesium, sodium, potassium, carnitine, bicarbonate, and water via supplementation or enteral feeding.

Caloric intake	100-110% RDA*
Protein	Normo- proteic for age (10-12% total calories)
Lipids	30% total calories
Carbohydrates	60% total calories
Micronutrients	Adequate
Salt and fluid supplementation	As needed

* *No evidence indicates that caloric intake in excess to 150%RDA is useful.*

Dietary reccommendations

- «4 P, kid friendly»

Pizza, pickles, pretzels and potato chips.

- Low acceptance for sweets, so it is difficult to use dietary supplements;
- Increase caloric density: unsatured fats, carbohydrate or lipid modules (to limit protein calories).
- Spicy and sour flavor foods, lemon and pepper.

Enteral nutrition

When oral feeding does not meet the nutritional need of the patient, enteral therapy is suggested.

- **Enteral support** with special formula.

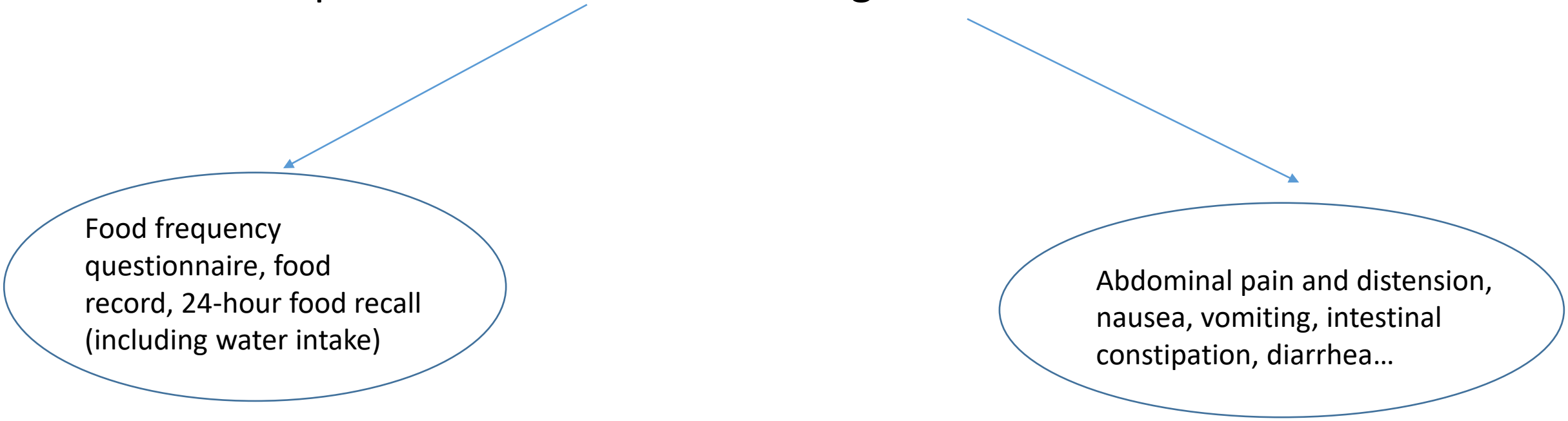
Or

- Completing meal by tube during the day.

Nutritional therapy should be **individualized**: encourage oral intake and estabilish tube feeding formula, volume and infusion time according to patient tolerance.

Enteral nutrition

- **Growth deficiency** in cystinosis patients is usually higher: optimizing weight gain.
  - The caloric and protein intake is calculated according to the RDA: optimal food evaluation and gastrointestinal evaluation.



Food frequency questionnaire

	Categoria di cibi	Quanto spesso consuma i seguenti prodotti?	E qual è la porzione media al giorno?	Esempi di porzioni
1	Pane dolce (pane zuccherato, pane con uvetta...)	○ Mai o meno di una volta al mese ○ 1-3 volte al mese ○ 1 volte a settimana ○ 2-4 volte a settimana ○ 5-6 volte a settimana ○ tutti i giorni	○ 40 g o meno ○ tra 40 g e 120 g ○ più di 120 g	Una fetta di pane grande=30 g Una fetta di pane piccolo=20 g
2	Pane/Fette biscottate/Panino croccante/pane francese/wafer di riso	○ Mai o meno di una volta al mese ○ 1-3 volte al mese ○ 1 volte a settimana ○ 2-4 volte a settimana ○ 5-6 volte a settimana ○ tutti i giorni	○ 40 g o meno ○ tra 40 g e 120 g ○ più di 120 g	Una fetta di pane grande=30 g Una fetta di pane piccolo=20 g 1 fetta biscottata=10 g 1 panino croccante=40 g
3	Latte zuccherato (esempio: latte aromatizzato, latte e cioccolato, porridge, Latticello zuccherato...)	○ Mai o meno di una volta al mese ○ 1-3 volte al mese ○ 1 volte a settimana ○ 2-4 volte a settimana ○ 5-6 volte a settimana ○ tutti i giorni	○ 200 ml o meno ○ tra 200 ml e 400 ml ○ tra 400 ml e 600 ml ○ 600 ml e più	1 tazza=225 ml 1 bicchiere= 150 ml 1 cartone= 200 ml 1 coppa= 125 ml 1 ciotola=250 ml
4	Latte non aromatizzato e senza aggiunta di zucchero	○ Mai o meno di una volta al mese ○ 1-3 volte al mese ○ 1 volte a settimana ○ 2-4 volte a settimana ○ 5-6 volte a settimana ○ tutti i giorni	○ 200 ml o meno ○ tra 200 ml e 400 ml ○ tra 400 ml e 600 ml ○ 600 ml e più	1 tazza=225 ml 1 bicchiere= 150 ml
5	Caffè e te senza zucchero	○ Mai o meno di una volta al mese ○ 1-3 volte al mese ○ 1 volte a settimana ○ 2-4 volte a settimana ○ 5-6 volte a settimana ○ tutti i giorni	○ 200 ml o meno ○ tra 200 ml e 400 ml ○ tra 400 ml e 600 ml ○ 600 ml e più	1 tazza= 125 ml 1 tazza=225 ml
6	Caffè e te con zucchero	○ Mai o meno di una volta al mese ○ 1-3 volte al mese ○ 1 volte a settimana ○ 2-4 volte a settimana ○ 5-6 volte a settimana ○ tutti i giorni	○ 200 ml o meno ○ tra 200 ml e 400 ml ○ tra 400 ml e 600 ml ○ 600 ml e più	1 tazza= 125 ml 1 tazza=225 ml
7	Succo di frutta	○ Mai o meno di una volta al mese ○ 1-3 volte al mese	○ 200 ml o meno ○ tra 200 ml e 400 ml	1 cartone=200 ml 1 tazza= 125 ml

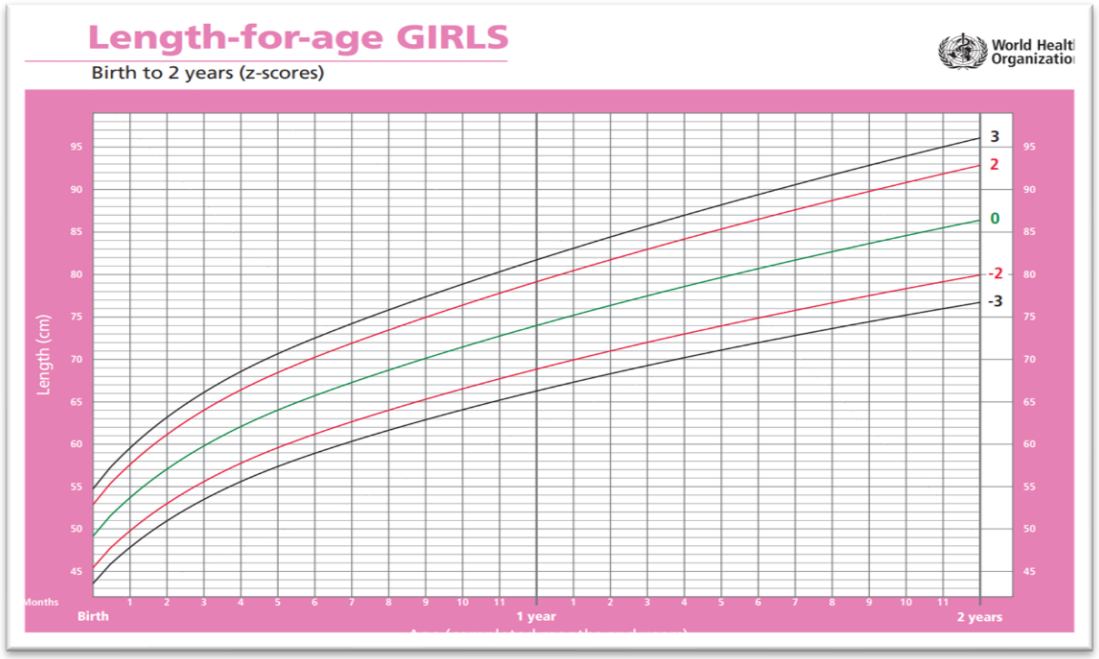
Antropometric evaluation

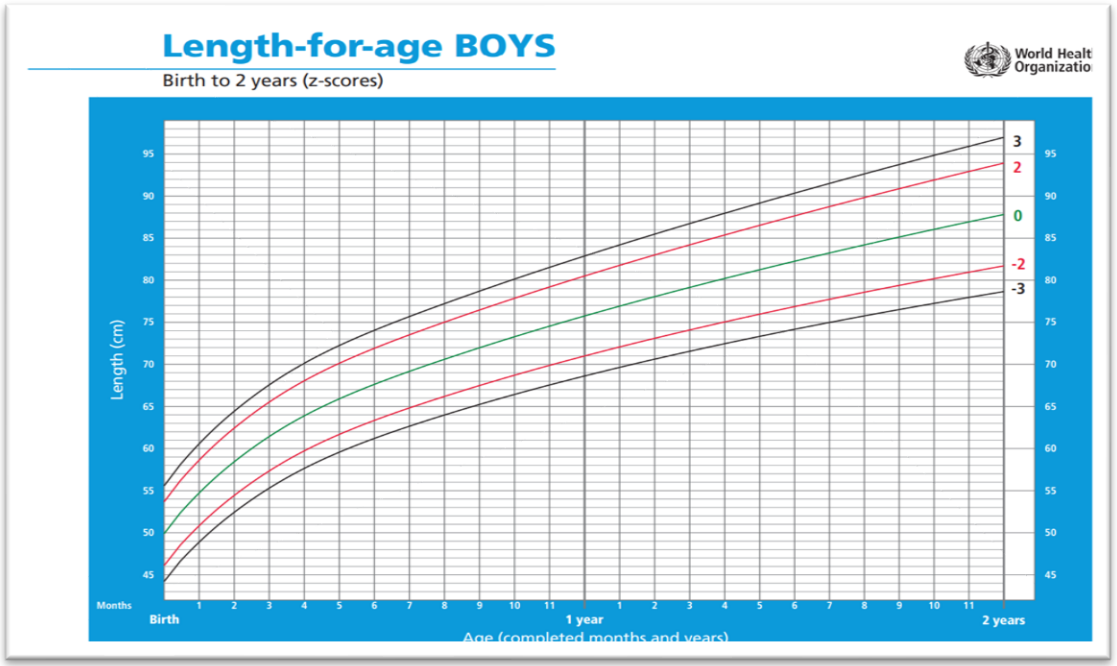
- Weight, height, head circumference.

Antropometric evaluation

- With the weight and stature data, it is possible to evaluate the indices:

Weight for Height, Height for Age, Weight for Age and Body Mass Index (BMI) for age, with percentile or z-score evaluation according to World Health Organization recomendations.





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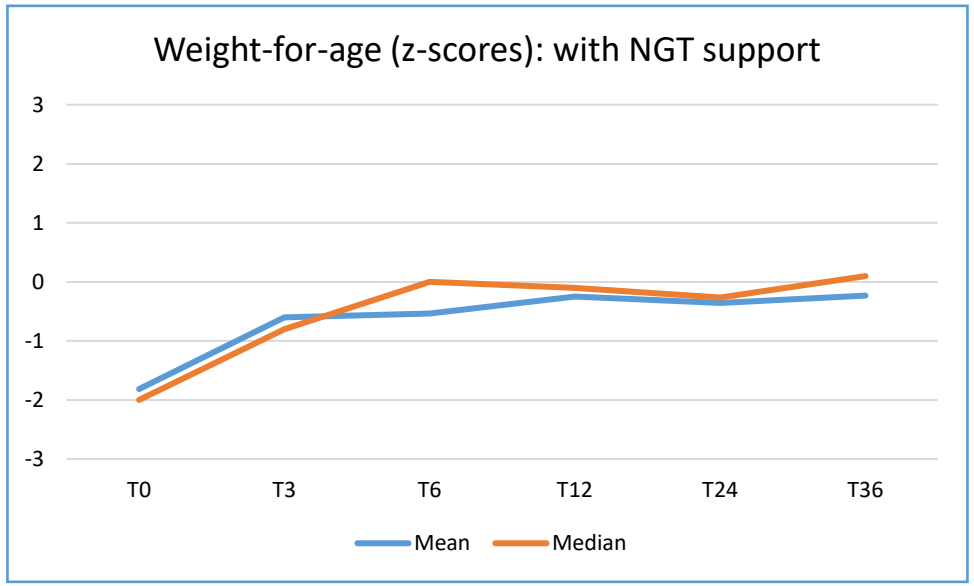
Patient	Gender	Age at diagnosis of cystinosis (years)	Age Enteral feeding commenced (years)	Duration of enteral feeding (years)	Overnight /daily Enteral formula	Caloric supplement powder
1	F	1	1	2,11	Formula milk	Maltodextrin
2	F	0,8	0,8	2	Formula milk+ Aproteic milk	Glyco lipid supplement
3	F	1,5	1,5	2	Complete formula	Glyco lipid supplement
4	F	1,7	1,7	1,1	Growth milk	NA
5	M	0,9	0,9	In progress	Formula milk + Aproteic milk	NA
6	M	0,7	0,7	2,1	Formula milk	Glyco lipid supplement
7	M	1,5	1,5	2,11	Complete formula	Glyco lipid supplement
8	M	1,4	1,4	2,9	Formula milk	Maltodextrin
9	M	1,1	-	-	-	-
10	M	1,11	-	-	-	-
11	M	5,5	-	-	-	-
12	F	1,7	-	-	-	-
13	F	0,11	-	-	-	-
14	M	1,9	-	-	-	-

Patients with enteral support (NGT)	Mean % RDA protein intake during follow up with enteral support	Mean % RDA caloric intake during follow up with enteral support	% RDA Protein intake (nasogastric tube removal)	% RDA caloric intake (nasogastric tube removal)
1	155	89	165	96
2	163	152	300	94
3	150	82	183	100
4	175	111	200	150
5	141	114	In enteral support	In enteral support
6	135	89	266	91
7	150	100	307	73
8	218	107	200	99

Patients with no enteral support	Mean % RDA protein intake during follow up	Mean % RDA caloric intake during follow up
9	247	80
10	169	60
11	200	83
12	152	89
13	190	90
14	183	89

Auxological parameters



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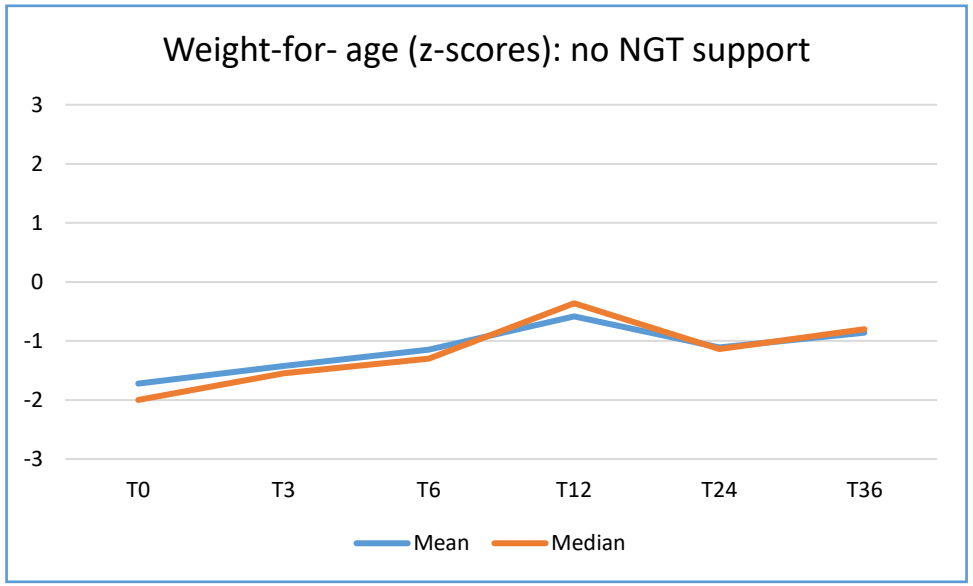

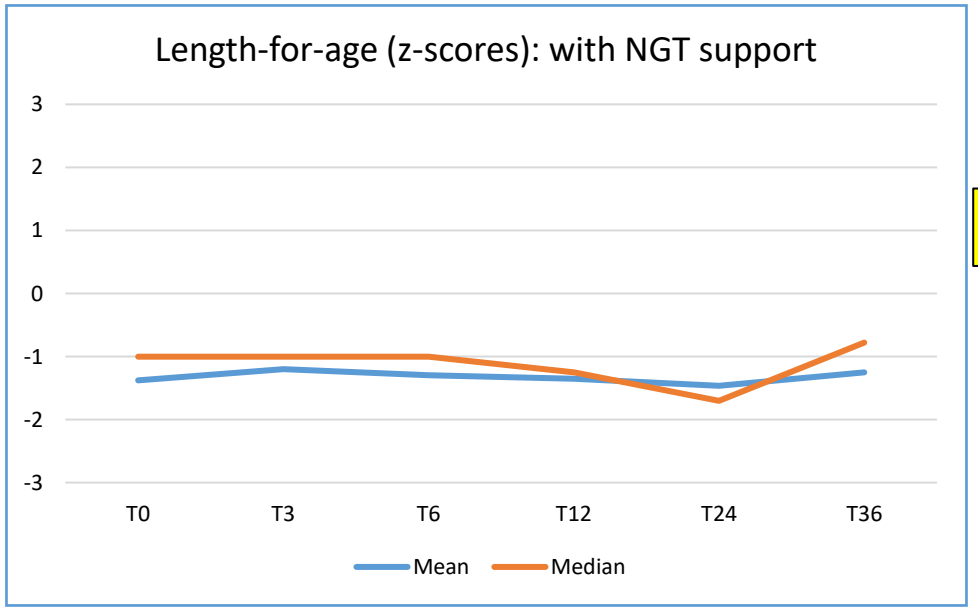

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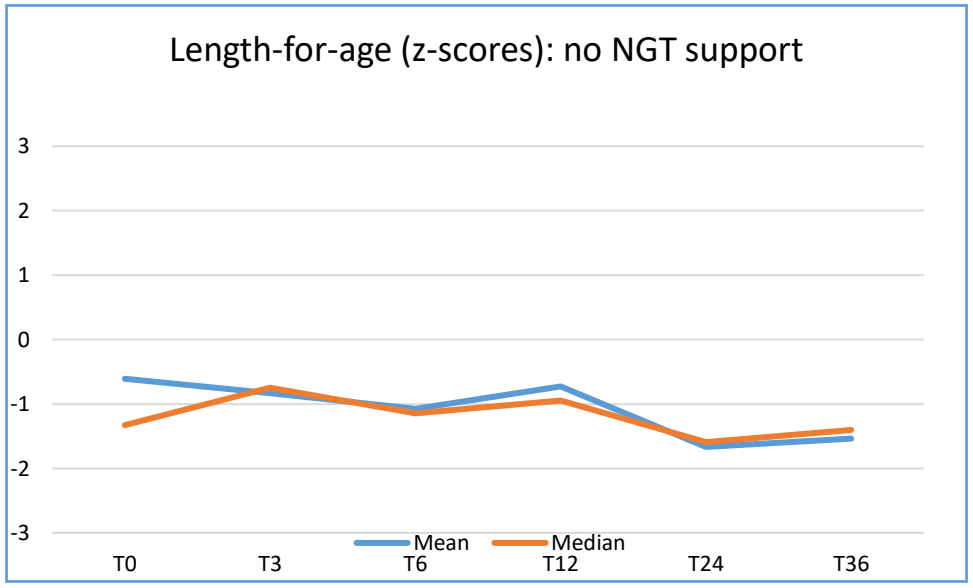
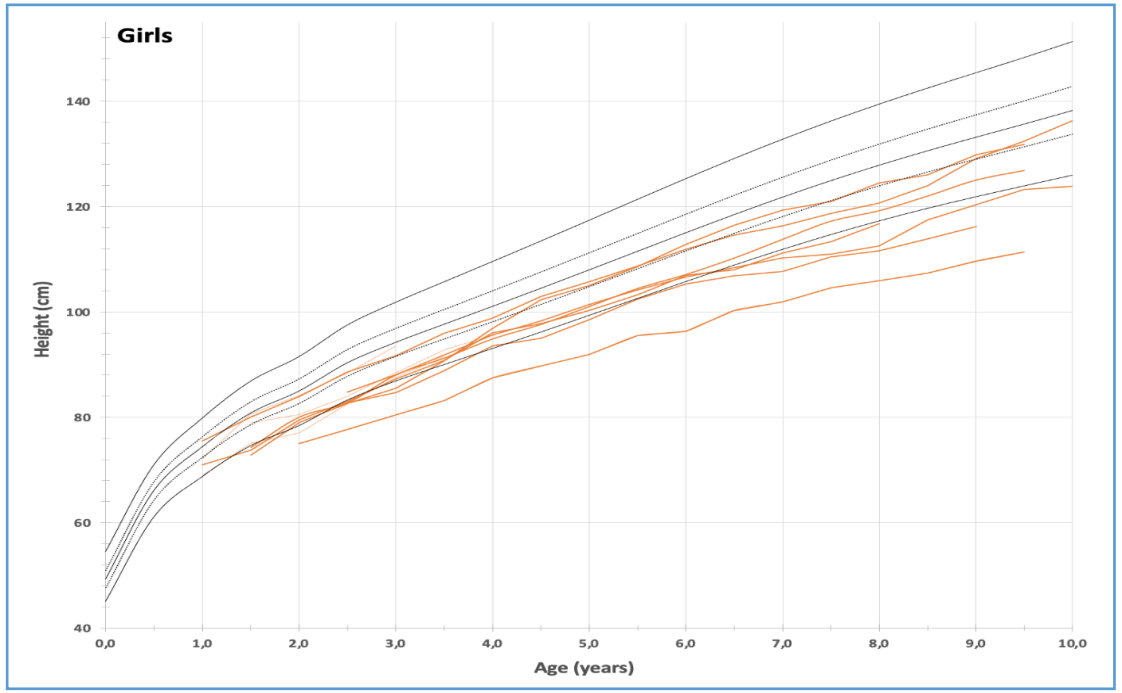
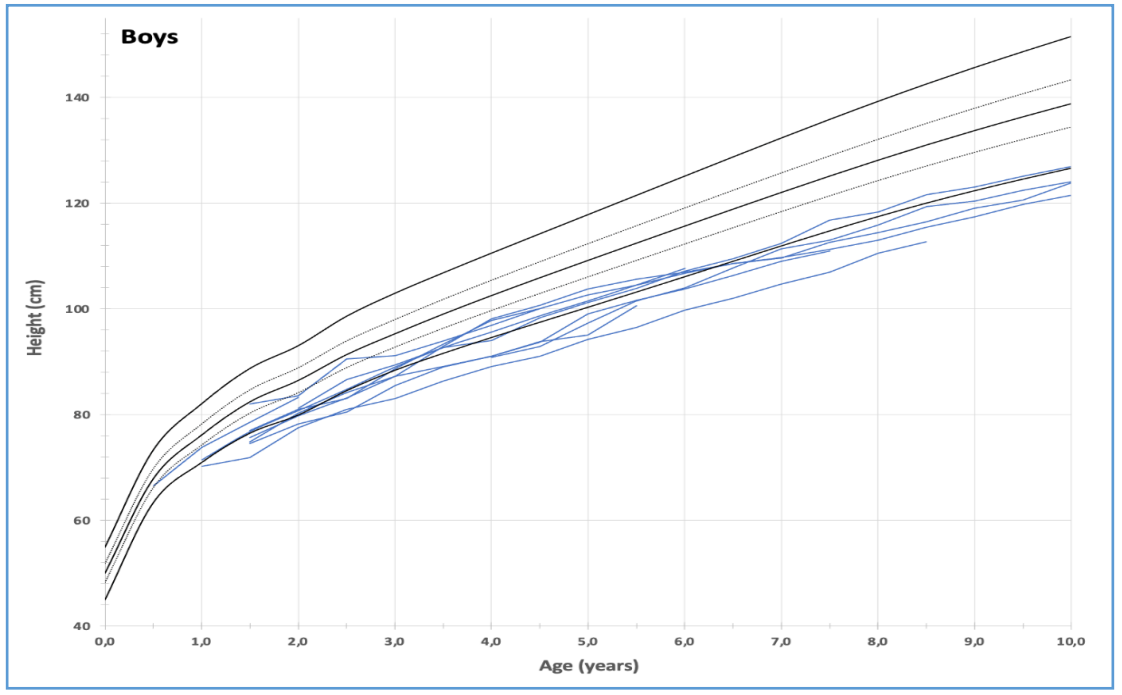


T0: TIME AT DIAGNOSIS
T3: 3 MONTHS FROM DIAGNOSIS
T6: 6 MONTHS FROM DIAGNOSIS
T12: 1 YEAR FROM DIAGNOSIS
T24: 2 YEARS FROM DIAGNOSIS
T36: 3 YEARS FROM DIAGNOSIS

21 Patients with cystinosis in our Nephrology Unit followed in 10 years (enteral & no enteral support):
Growth curves (Height)

Small variations in the height/weight status reflects the relatively long period of poor growth before the diagnosis.





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Renal Dietitian roles in cystinosis patients

Renal dietitians are required members of the interdisciplinary team who care for cystinosis patients.

Assessment:

- **Dietary intake assessment** with diet interviews and/or diaries.
- Anthropometry .
- Check **grow chart**.
- Physical assessment.
- Reccommending supplements for inadequate protein and/or energy intake.
- Nutrition counseling on protein needs, on electrolyte and/or mineral intake
- Nutrition counseling on weight (increasing/decreasing calories)
- Nutrition counseling: other (eg appetite, other comorbidities).
- Solid **supportive nutrition plan**.
- **Monthly** dietetic review (for infants in the first year of life) to prevent inadequate caloric intake.
- Ongoing **email communication** with families to modify, if necessary, dietetic plan.

Take- home message

- Dietetic management involves providing nutritional support via supplements or in most cases tube feeding, along with **growth monitoring** and helping parents to manage **feeding difficulties**.
- Organize dietetic assessment with parent or carer present.
- Plot anthropometry on growth charts.
- Assess **energy** requirements. If catch up growth is required, use 130-150 kcal/kg (infant) or 120% estimated average requirement for energy.
- Estimate **protein** requirements (RDA for age) and allow extra for catch up growth if necessary (to achieve protein energy ratio of 11-12%)
- Assess tolerance.

Nutritional management of children with cystinosis. Clinical guideline- University Hospitals Bristol

THANKS FOR YOUR ATTENTION