ADOLESCENT WITH CF, LIVER DISEASE, RENAL FAILURE AND POOR GROWTH

ECFS 2022, Cystic Fibrosis Nutrition Group Meeting

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OVERVIEW

- Case
- Growth
- Nutrional intake
- Conclusions and Future consideraties

CASE

13 years old adolescent girl

Medical historie

- CF homozygoot dF508, 2018 Symkevi (stopped in 2020), februari 2022 Kaftrio
- Pancreasinsufficient: Pancrease enzymes Creon 10.000, vit ADEK
- Milde aortic valve abnormality
- 2013 small for gestational age (SGA) and partial Growth Hormone (GH) deficiency, useing growth hormones had insufficient effect. SGA probebly causes by chronical illness/malnutrition.
- 2017 Hypertension and a progressive Chronic Kidney Disease (CKD) based on nephronopthisis (NPHP 1x missing, 1x mutation): Amlopidine and potassium supletion.
- 2019 CF related diabetic: Levemir (daily dose)
- 2020 increased liverenzymes most likely Drug-Induced Liver injury (DILI) caused by Symkevi
- No signs of puberty
- Before Kaftrio multiple admissions to the hospital

What is the goal for growth?

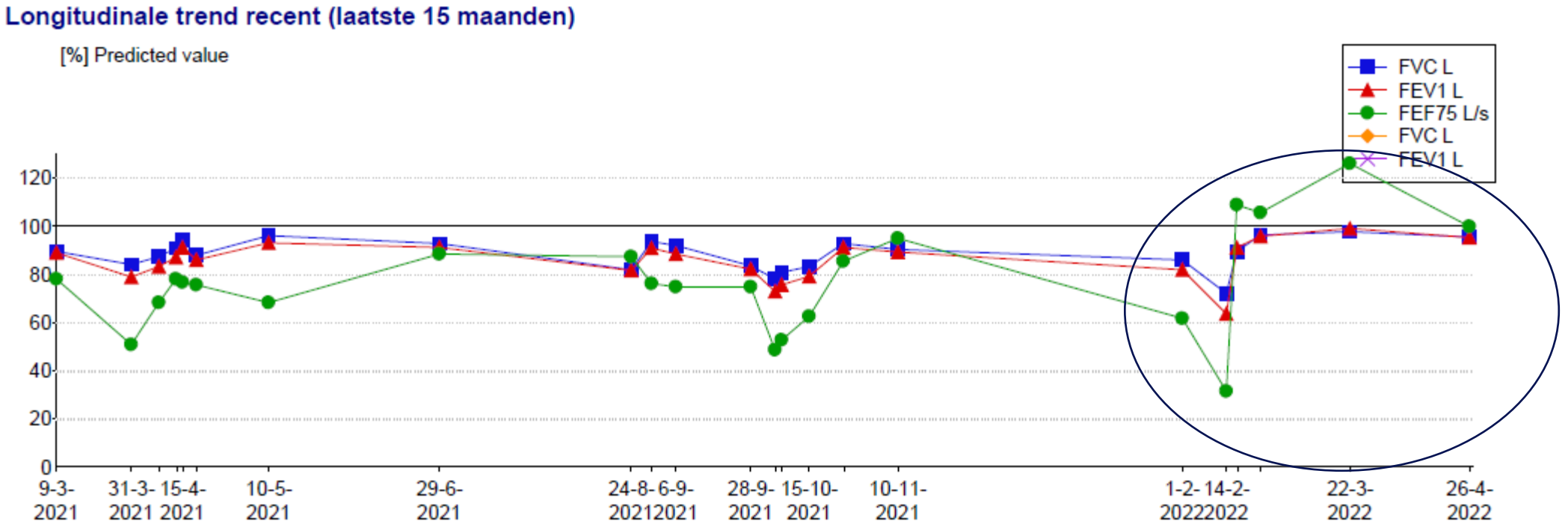
What to do with her diet?

CASE

13 years old adolescent girl

Longfunction

- FEV1 increased from 63.6% tot 95.2% in April
- Increase of 99% was with 1 daily dosis of Kaftrio
- Decreased tot 95% with 3 times a week Kaftrio



Szczesniak, R., Heltshe, S. L., Stanojevic, S., & Mayer-Hamblett, N. (2017). Use of FEV_1 in cystic fibrosis epidemiologic studies and clinical trials: A statistical perspective for the clinical researcher. *Journal of cystic fibrosis : official journal of the European Cystic Fibrosis Society*, *16*(3), 318–326. https://doi.org/10.1016/j.jcf.2017.01.002

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CASE

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Relevant bloodwork

Urea	Elevated 12.4 mmol/L (3,3-5,6)
Creatinine	Elevated 222 umol/L (31-68)
Schwartz eGFR	Low 23 mL/min (>90)
Sodium	Normal (with supplements)
Potasium	Normal (with supplement)
Phosphaat	Elevated (before dietary intervention) highest 1,94 mmol/L
ASAT/ALAT/GGT	Elevated
Vit B12	No deficiency

CASE

13 years old adolescent girl

Anthropometry

	Before Kaftrio	After Kaftrio
Weight (SD)	31,8kg	34,5kg
Weight for length	0,61 SD	1,09 SD
Length	-3,9 SD	-3,85 SD
BMI	-0,7 SD	-0,14 SD
Bone mineral density (2019 vs 2022)	-1,7 SD from Z-score for age and gender	-3,3 SD from Z-score for age and gender

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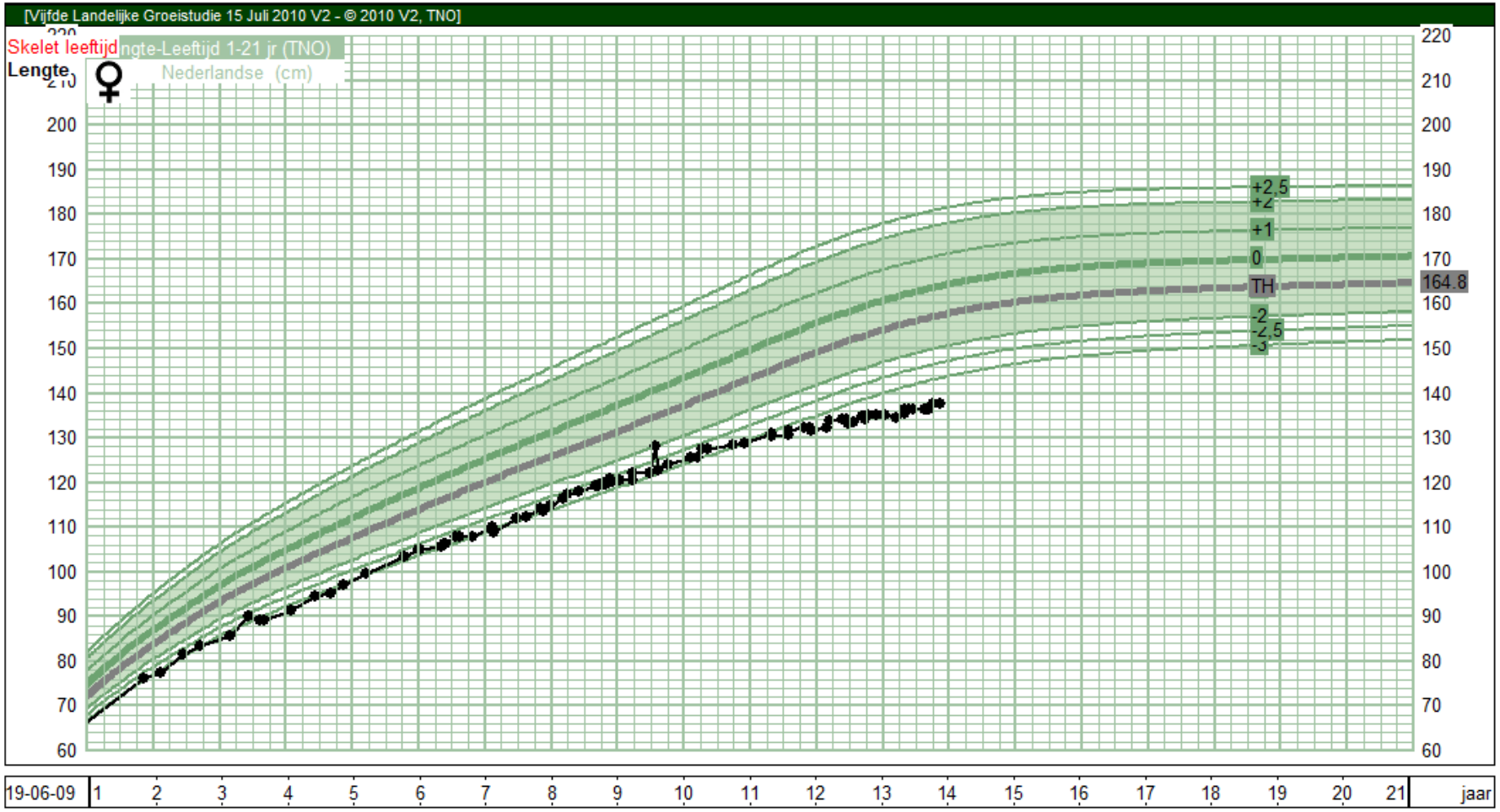
GROWTH

Length-age



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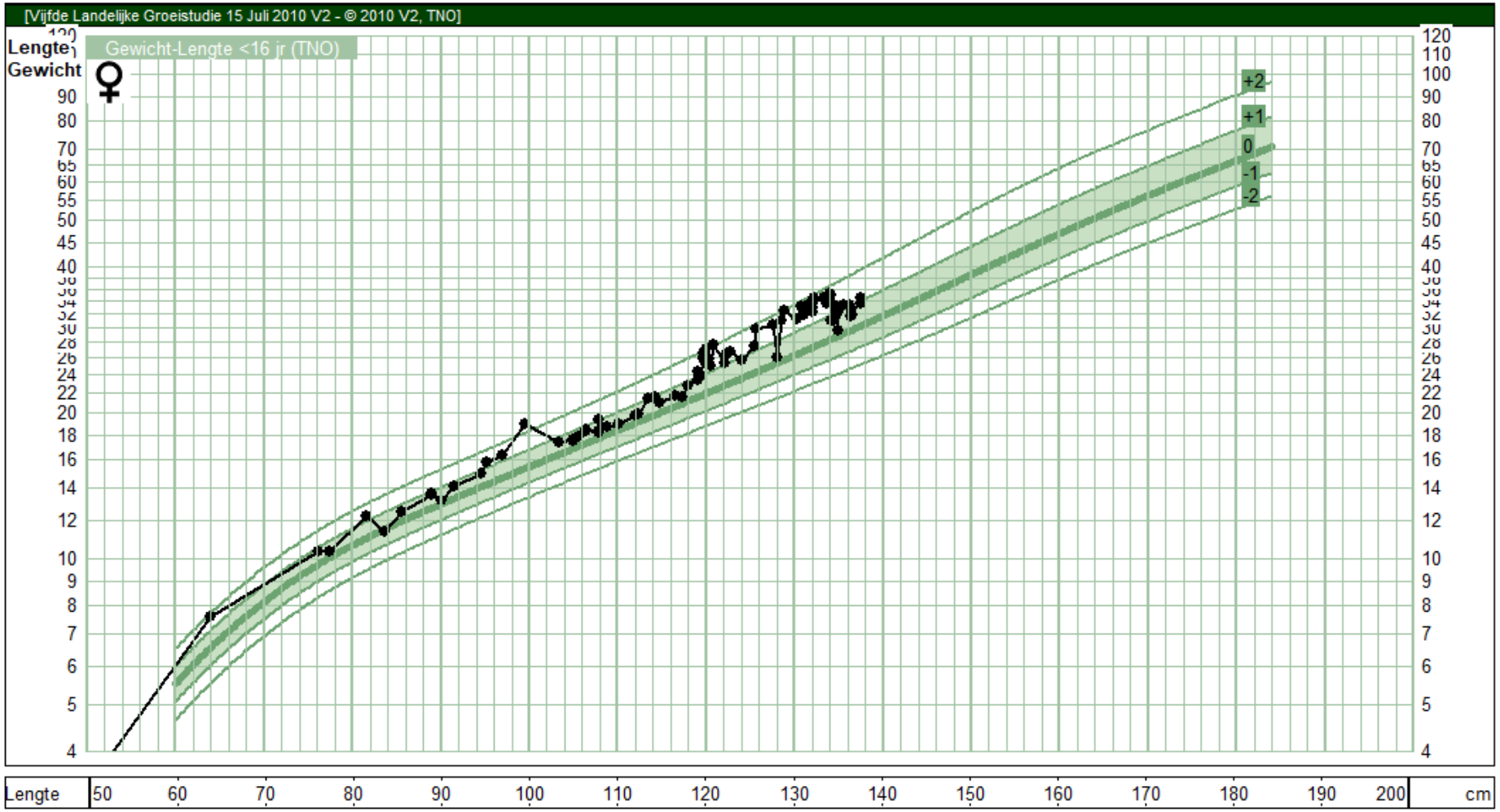
GROWTH

Weight-length



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WHAT IS THE GOAL FOR HER GROWTH

ESPEN-ESPGHAN-ECFS guidelines on nutrition care for infants, children, and adults with CF

- Children 2-18 years the target is a body max index (BMI) at or above the 50th percentile (or 0SD) for healthy children

Turck, D., Braegger, C. P., Colombo, C., Declercq, D., Morton, A., Pancheva, R., Robberecht, E., Stern, M., Strandvik, B., Wolfe, S., Schneider, S. M., & Wilschanski, M. (2016). ESPEN-ESPGHAN-ECFS guidelines on nutrition care for infants, children, and adults with cystic fibrosis. *Clinical nutrition (Edinburgh, Scotland)*, *35*(3), 557–577. https://doi.org/10.1016/j.clnu.2016.03.004

CASE

13 years old adolescent girl

24h recall

- Breakfast: skips
- Snack: -
- Lunch: 1 slice of bread with butter and cheese spread and cookie
- Snacks: -
- Diner: Puree with vegetables, meat and gravy
- Dessert: Dubbelvla (dairy), snack-a-jack or chips
- Drinken: fristi (sweetened diary drink), Optimel (sweetend (light) diary drink), limonade and cola.

Intake low in calories, protein en vitamines and minerals. But high in phosphate!!

Dietary intervention

- Replace cheese spread and cola for other options
- Restart Oral Nutritional supplement (special for kidneys Nepro ®)
- 3 times a day a snack
- Change suger right drinks for light versions or water

CASE

13 years old adolescent girl

- More appetite sinds start Kafrtio, more energy
- Stool: 1-2 times a day. Says its not greasy
- Creon compliance: forgets with snacks and sometimes before diner. Eats alone a lott of the times
- Social: 4 children who lives at home, 3 adult children don’t live at home any more. Sister with DM type 1. Multiple social problems. Simple explanation is important

SUMMARY

- High in sugar rich beverages
- Dietary restrictions because of CFRD and renal problems
- Poor creon compliance
- Limited intake of specifick food groups
- Lack of support during diner times
- Hard to instruate about healthy food chooses

DIETARY RECCOMONADATIONS

ESPEN-ESPGHAN-ECFS guidelines on nutrition care for infants, children, and adults with CF

- High caloric (110-200%) and high-fat diet (30-40 EN%) + pancrease enzymes and fat-soluble viatmines
- Increased protein intake (20 EN%)

Disadvantages of high fat diets and high caloric diets

- Increase in saturated fats and trans fatty acid → cardiovasculaire disease

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CONCLUSION

Mulitple factors to take into account

- Different nutrition needs: CF, growth deficiency and renal problems
- Difficult to estimate growth goal
- IQ and social situation

Future

- Weekly doses of Kaftrio: effect on appetite and growth?
- Start multivitamine supplement (compliance?) or tube feeding
- Nutrional assesment

LITERATURE

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THANK YOU

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