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**Use of modified cornstarch in patients  
affected by GSD I:  
an observational in-patient study**

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## Patients and methods

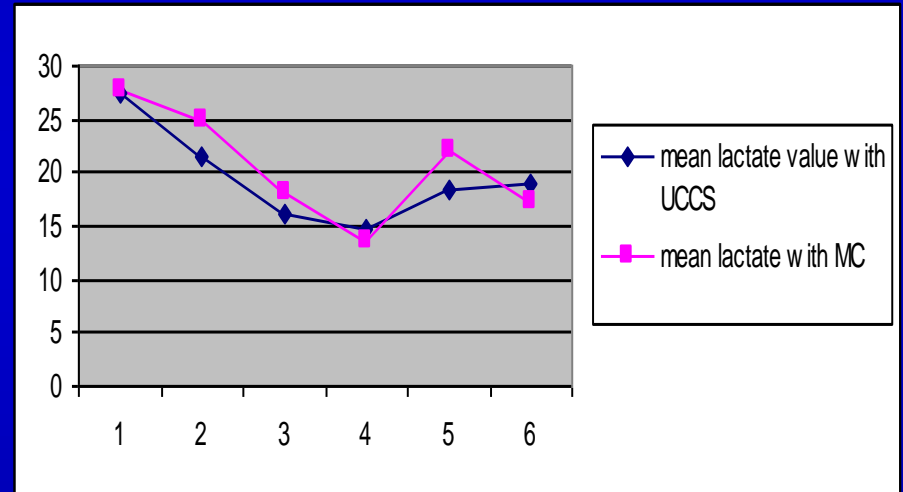
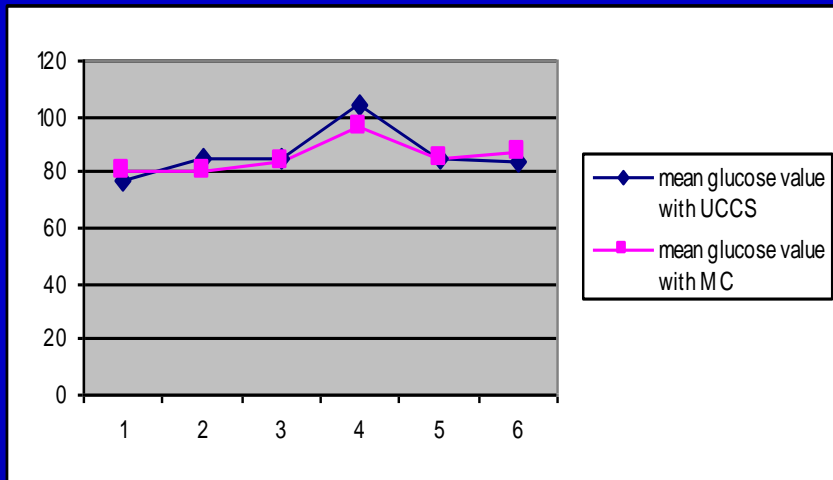
- 6 patients with GSD I (1 Ia and 5 Ib) were recruited from our Metabolic Unit and the challenge with modified cornstarch was proposed.
- The diagnosis of GSD I was based on a liver biopsy showing reduced activity of the appropriate enzyme or on a mutation in the appropriate gene.

# Patients and methods

- The study was a consecutive, 48 hours, in-patient stay at San Paolo Hospital. Patients continued their usual diet, keeping their same time meals as at home.
- Glucose and lactate were measured at pre-prandial time (5 or 6 times a day).
- The usual starch (UCCS) was administered on the first 24 hours, the alternative (MC) on second day of the study. We decided, for safety reasons, to administer the same dose of MC as dose of UCCS provided in each patient diet.
- We collected patient's feelings (about taste, texture, digestibility) and signs or symptoms (like meteorism, swelling sensation, diarrhea).

# Results and discussion

- We performed a statistical analysis using Wilcoxon test: we confronted mean levels of glucose and lactate using UCCS and MC, in each time of their 24-hours profile.
- There was no statistically significant difference.



- We calculated also the variability of glucose and lactate levels in the whole cohort, with UCCS and MC: there was no statistically significant difference in using MC

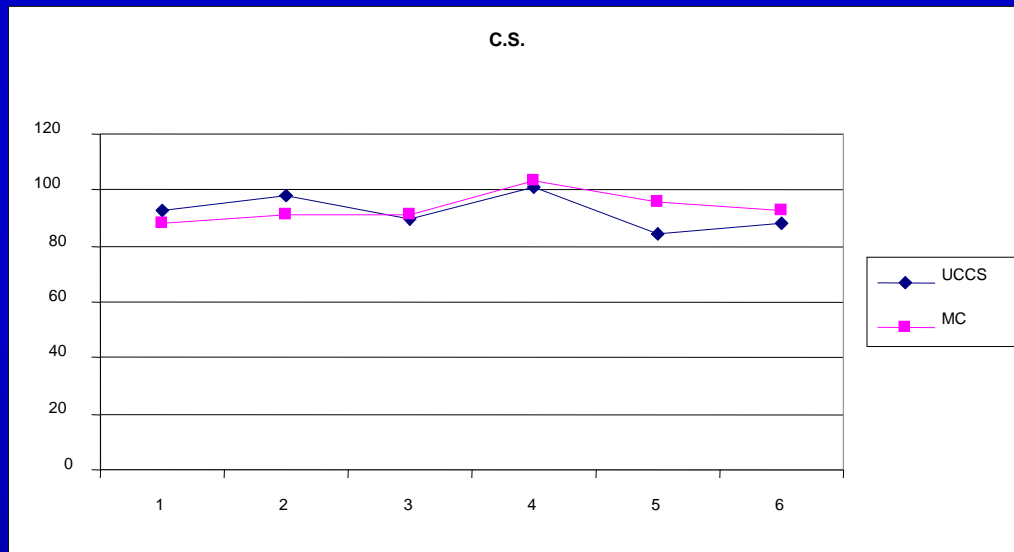
# Results and discussion

## Feelings and symptoms

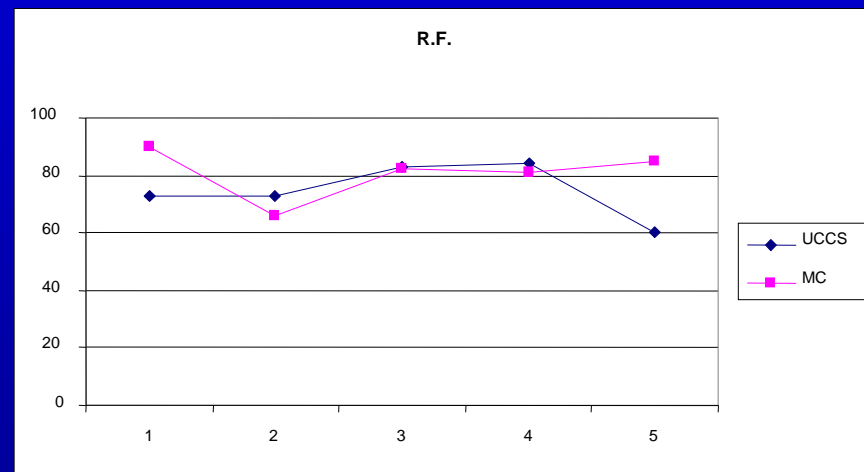
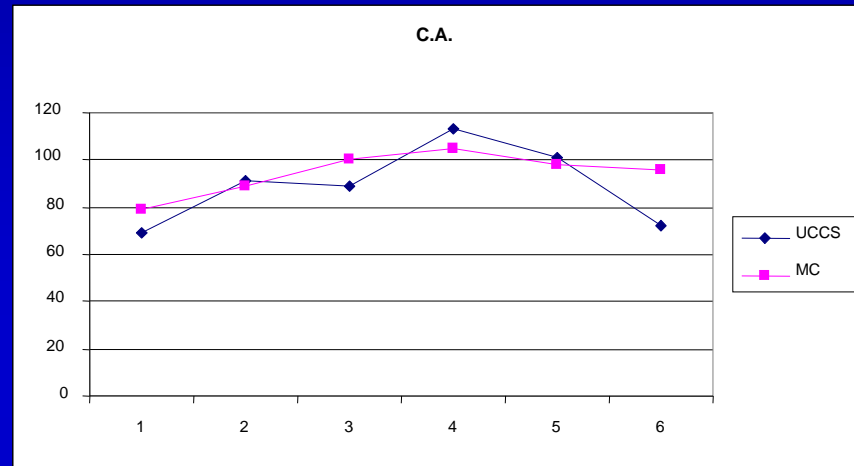
	BF	CA	CS	BS	RF	MC
patient's feelings	↓apetite	bad taste bad texture	none	none	bad taste bad texture	none
signs/symptoms	↓meteorism ↓swelling	↑meteorism ↑swelling	none	none	↓meteorism ↓swelling	none

# Results and discussion

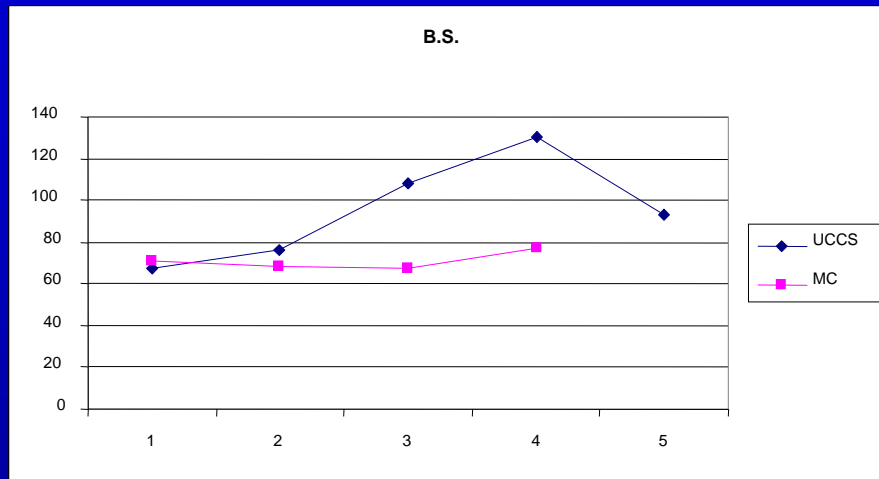
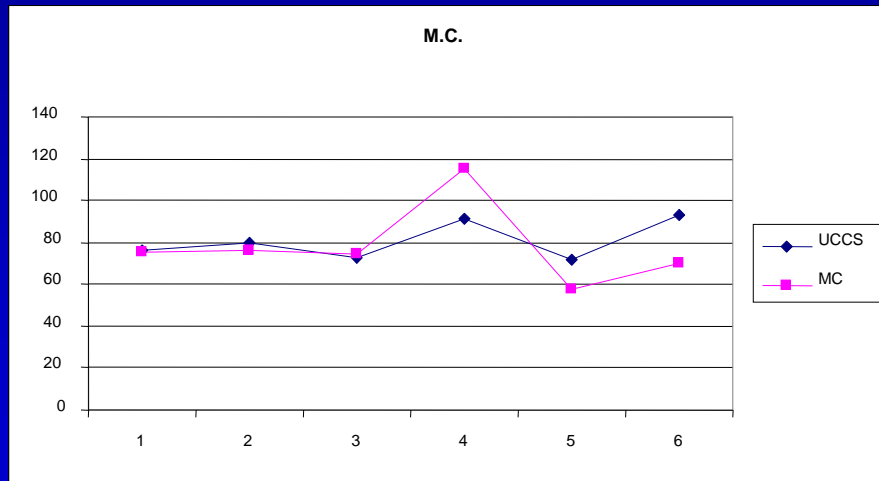
No difference



- Small biochemical advantage
- Bad texture and taste

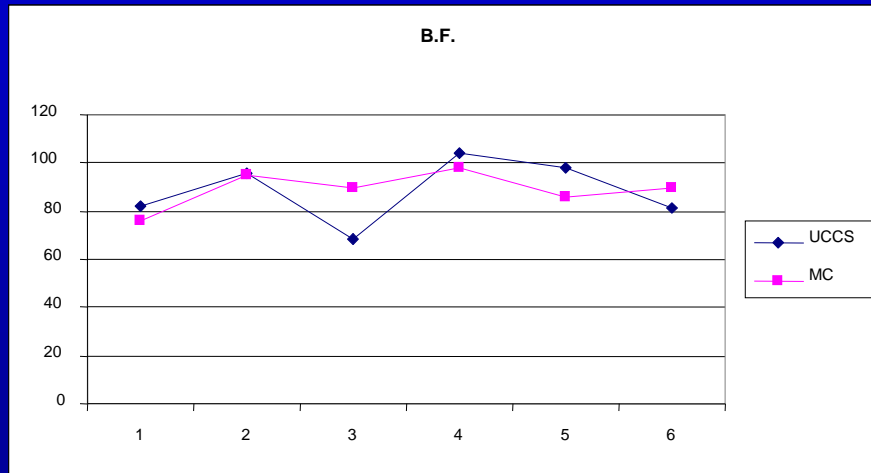


# No advantage



# Results and discussion

- Despite the inconsistency of statistical analysis, MC maintained more constant levels of glucose and lactate compared to UCCS in one of our patient.
- We tried the administration of MC for a longer period; we recommended the patient to continue measurements of glucose-meter before all meals also at home.
- After 5 months we noted a clinical improvement: B.F. has less prominent abdomen, liver size and echogenicity (steatosis) are stable, and parents refer that the child is less hungry and that the glucose-meter values are more constant.
- Laboratory analysis are stable after these first 5 months of treatment with MC (slight decrease of hepatic transaminases).



# Conclusions

- We studied a small number of heterogeneous patients with large differences in age, type and severity of disease, who often require meticulous individualized management.
- The newly created modified cornstarch could be a promising alternative to traditional cornstarch preparations.
- Further studies are necessary to examine the role of this novel starch as part of the standard dietary regimen of larger numbers of patients for a greater period of time.