

Duration of Fasting Studies Required to Demonstrate a Cure for Patients with Focal Congenital Hyperinsulinism.

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Introduction

Focal disease of the pancreas causing hyperinsulinism (FHI) has 2 essential characteristics 1) the inheritance of a paternally derived mutation in *ABCC8* or *KCNJ11* in conjunction with loss of a region of the maternal chromosome 11p13.1 and 2) the ability to be cured by resection of the focal lesion while sparing the majority of the pancreas. At the time of surgery the surgeon resects the focal lesion until clear margins are available. We perform a fasting study postoperatively to determine if the patient is cured or not. In our multidisciplinary team approach we terminate our fasting studies when either a plasma glucose is < 2.8 mmol/L, BOHB is > 1.7 mmol/L or a max fasting duration appropriate for the patient's age has been reached. There is current debate as to how long one should fast an infant to demonstrate cure of FHI.

Aim

The aim of this study is to demonstrate the length of time to either hypoglycemia < 2.8 mmol/L or BOHB > 1.7 mmol/L in order to prove cure in patients with FHI in whom surgery has been deemed successful at the time of frozen section.

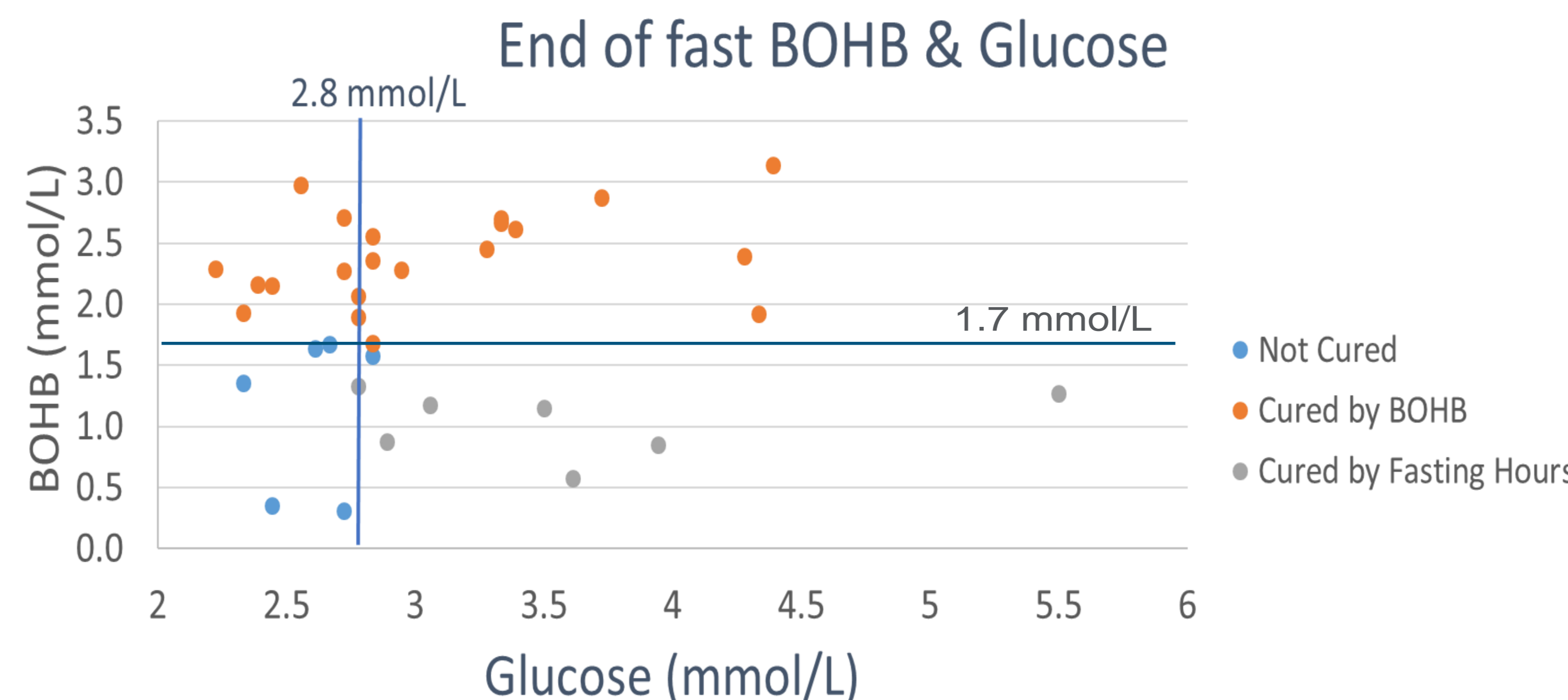
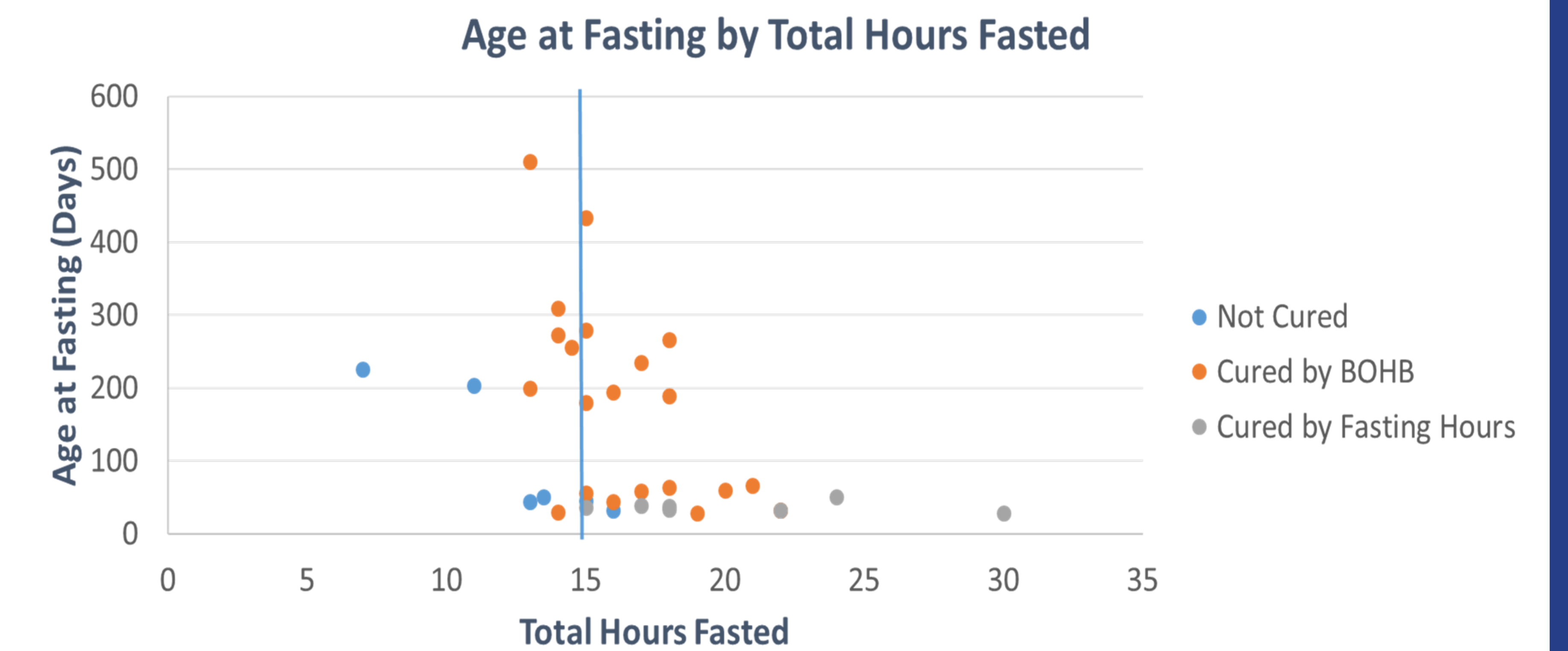
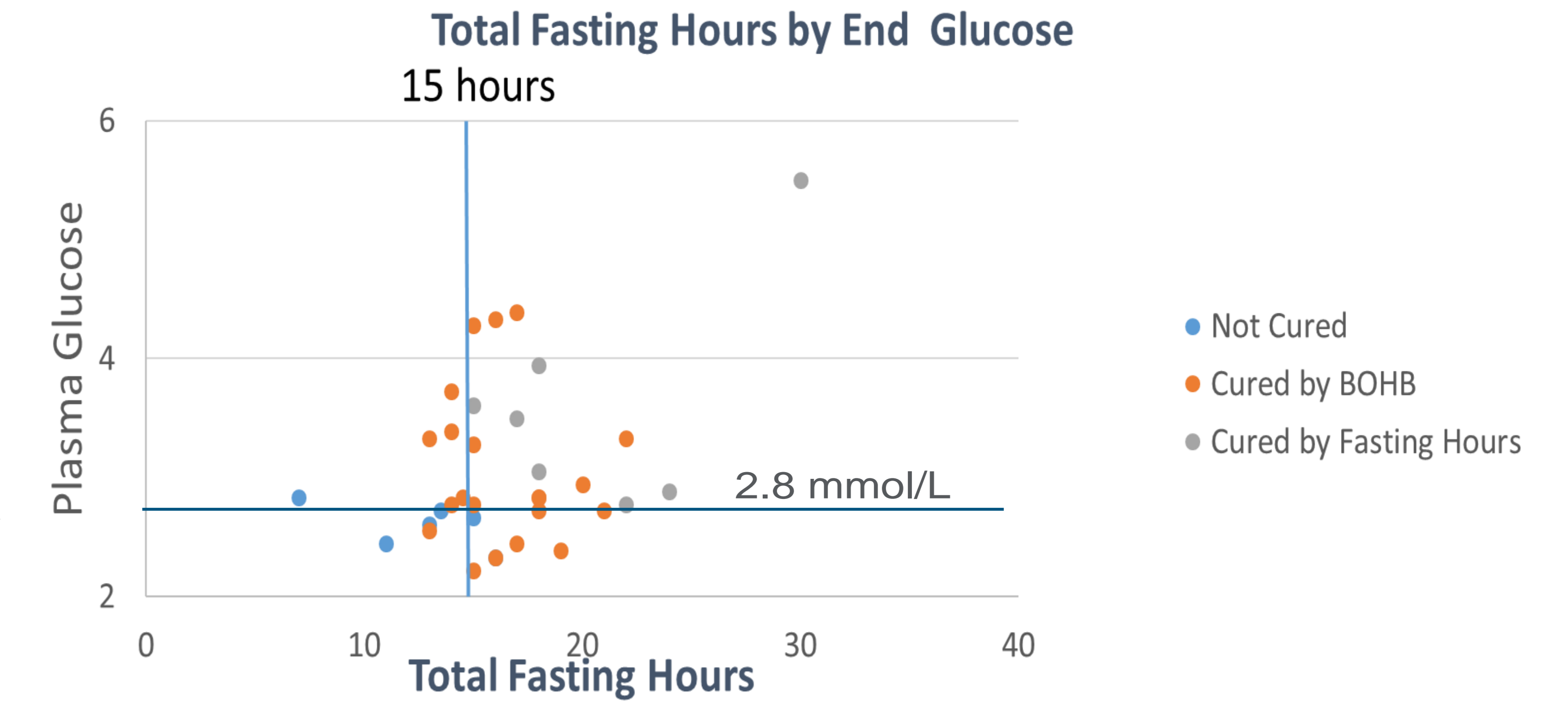
Methods

We conducted a retrospective analysis on 34 patients with focal disease. Patients were divided into 3 groups. Group 1: those not cured using the a priori criteria, Group 2 those cured and fasted until glucose < 2.8 mmol/L or BOHB > 1.7 mmol/L, group 3 those cured and fasted based on time.

Results

See Table 1 for fasting details. In Group 1, 5 subjects fasted more than 9 hours and 4 subjects fasting > 12 hours. In Group 2, 7/21 subject fasted 18 hours. All subjects in Group 3 fasted > 18 hours. Of the patients determined cured (Groups 2 and 3), the 9 hour glucose was above 3.88 mmol/L (70 mg/dL) in 25/28 patients, whereas in Group 1 (not cured) the glucose was > 3.88 mmol/L in only one subject. In our case an average length of stay for cured focal patients is 22.1 days.

	Group 1	Group 2	Group 3
N	6	21	7
Fasting (hours)			
Median	13.25	16	18
Range	7-16	13-22	15-30
Age (days)			
Median	48.5	189	36
Range	33-226	28-511	28-51
LOS (days)			
Median	27	23	20
Range	24-46	8-38	15-25
Gender (n)			
Female	3	6	3
Male	3	15	4
Paternal mutation in KATP channel gene only	6	18	6



Conclusion

We recommend that fasting studies to demonstrate cure in patients with FHI be performed until either the glucose is < 2.8 mmol/L and the beta > 1.7 mmol/L or 15 hours whichever comes first.



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