

Abnormal Glucose Tolerance in Infants and Young Children with Cystic Fibrosis

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ONLINE DATA SUPPLEMENT

Supplemental methods:

The antibody pairs in the panel were specific only to the desired analyte and exhibited no or negligible (<2%) cross-reactivity with other analytes in the panel. The minimal detectable concentration for the insulin assay was 14.9 pmol/L and C-peptide the minimal detectable concentration was 0.33pmol/L. Coefficient of variation for the insulin assay was 1.4%; C-peptide, 2.7%; and glucose, 1.5%.

Supplemental table:

Supplemental Table: Genotype, glucose tolerance status and pancreatic status for subjects with cystic fibrosis

Subject number	Mutation1/Mutation 2	glucose tolerance category*	fecal elastase** (µg/g)	status of exocrine pancreas†
1	delta F508/G542X	NGT	<6	insufficient
2	Delta F 508/ 621+1G>T	NGT	<6	insufficient
3	delta F508/G551D	NGT	29	insufficient
4	delta F508/c.1807delG	could not be classified	21	insufficient
5	delta F508/G551D	IGT	89	insufficient
6	delta F508/G551D	NGT	53	insufficient
7	deltaF508/G542X no 5T	NGT	<6	insufficient
8	delta F508/delta F508	NGT	93	insufficient
9	delta F508/G542X†††	NGT	20	insufficient
10	delta F508/delta F508	NGT	NR	insufficient
11	delta F508/Q1313X	IGT	<15	insufficient
12	deltaF508/394delTT	IGT	NR	sufficient
13	delta F508/delta F508	NGT	NR	insufficient
14	delta F508/delta F508	NGT	98	insufficient
15	delta F508/delta F508	IGT	NR	insufficient
16	delta F508/delta F508	INDET	<50	insufficient
17	deltaF508/3120G>A	IGT	222	insufficient
18	delta F508/delta F508	DM	NR	insufficient
19	deltaF508/3120G>A	IGT	222	insufficient
20	delta F508/delta F508	IGT	NR	insufficient
21	delta F508/delta F508	NGT	57	insufficient
22	delta F508/ R553X	DM	21	insufficient
23	delta F508/delta F508	NGT	NR	insufficient
24	G551D / 621+1 G>T	NGT	NR	insufficient
34	delta F508/1154insTC	could not be classified	27	insufficient
35	delta F508/3849+10kbC>T††	could not be classified	7	insufficient
36	delta F508/delta F508***	could not be classified	17	insufficient

*NGT=normal glucose tolerance, IGT = impaired glucose tolerance, INDET= indeterminate glycemia, DM= diabetes mellitus. Please see text for definitions of these categories of abnormal glucose tolerance. Subjects listed as “could not be classified” did not have available data for all points and could not have official glucose tolerance status determined. The last three subjects had iv failure and had essentially no samples available. Fecal elastase results were abstracted from the clinical record and were performed at variable ages. **NR=not recorded in medical record. †Clinical pancreatic insufficiency or sufficiency was based on the classification recorded in the medical record by the child’s treating physician.††This subject’s genotype is typically associated with pancreatic sufficiency, but per the medical record the subject was classified as pancreatic insufficient. †††This subject has history of duodenal atresia s/p repair. ***This subject had history of meconium ileus.