

Online-Only Appendix A.

The CFRDT Study Group:

University of Minnesota, Minneapolis (PI-Antoinette Moran, MD)
Vanderbilt University, Nashville, Tennessee (PI-Bonnie Slovis, MD)
University of Pittsburgh, Pittsburgh, Pennsylvania (PI-Joseph Pilewski, MD)
St. Michael's Hospital, Toronto, Canada (PI-Elizabeth Tullis, MD)
University of Utah, Salt Lake City (PI-Theodore G. Liou, MD)
University of Vermont, Burlington (PI-Laurie Whittaker, MD)
Centre Hospitalier de l'Universite de Montreal, Canada (PI- Remi Rabassa Lhoret, MD)
St. Paul's Hospital, Vancouver, Canada (PI-Stuart Kreisman, MD)
Stanford University, Palo Alto, California (PI-Richard Moss, MD)
University of Cambridge, United Kingdom (PI-Amanda Adler, MD)
University of Florida, Gainesville (PI-Michael Stalvey, MD)
Children's Mercy Hospital, Kansas City, Missouri (PI-Kurt Midyett, MD)
Via Christi Specialty Clinic, Wichita, Kansas (PI- Daniel Doornbos, MD)

Online-Only Appendix B

Baseline characteristics of all patients enrolled, and by completion status.

	Total Cohort	Completed	Stopped Early	p-value [*]
Number	100	81	19 [§]	
OGTT status				0.57 ¹
CFRD FH-	74 (74%)	61 (82%)	13 (18%)	
IGT	26 (26%)	20 (77%)	6 (23%)	
Treatment Group				0.87 ²
Insulin	38 (38%)	30 (37%)	8 (42%)	
Repaglinide	31 (31%)	26 (32%)	6 (32%)	
Placebo	31 (31%)	25 (31%)	5 (26%)	
Female	47 (47%)	36 (44%)	11 (58%)	0.29 ²
Age (years)	27 ± 8	28 ± 1	25 ± 2	0.19 ³
BMI (kg/m2)	21.4 ± 3.0	21.4 ± 0.3	21.4 ± 0.7	0.99 ³
% Fat free mass	79 ± 10	79 ± 1	76 ± 2	0.23 ³
FVC %predicted	88 ± 22	89 ± 2	83 ± 5	0.27 ³
FEV1 %predicted	69 ± 25	69 ± 3	69 ± 6	0.93 ³
NIH score	82 ± 15	83 ± 2	77 ± 3	0.08
HbA1c (%)	6.0 ± 0.7	6.0 ± 0.1	6.0 ± 0.2	0.85 ³

Data for the total cohort are presented as mean ± SD; data for the sub-populations as mean ± SEM.

§ Among 19 stopping early, 8 were noncompliant/quit, 5 developed Fasting hyperglycemia, and 6 were stopped for other medical reasons.

* Tests for differences in baseline characteristics between those stopping early and those completing full study year.

¹ Fisher's Exact Test

² Chi-square Test

Online-Only Appendix C.

Reasons for Early Drop-Out in the 3 Arms (Insulin, Placebo, Repaglinide) of the CFRDT Trial.

Reason	Insulin	Placebo	Repaglinide	Total
Fasting Hyperglycemia	2	3	0	5
Other Medical*	1	1	4	6
Patient Quit	5	2	1	8
Total	8	6	5	19

*Other medical included chronic steroids (1 insulin, 1 repaglinide), pregnancy (1 repaglinide), rash attributed by patient to medication (placebo), patient chronically hospitalized and too ill to take medication (2 repaglinide). Fisher's Exact Test p=0.12 for association of reason for discontinuation with treatment.