

Online-Only Supplements

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E-Methods

Agreement on measures of guideline adherence

Clinicians may disagree regarding practice definitions that measure adherence to a guideline. To address potential disagreement, we conducted a survey collected and managed using REDCap electronic data capture tools hosted at University of Washington^{1,2} to obtain input on the appropriateness of a broad number of nutrition guideline measures with physicians and advanced practice providers. We surveyed 16 CF pediatric or joint pediatric and adult programs who participate in the CF Learning Network (CFLN) community. The CFLN is a collaborative group of CFF-accredited US Care Centers led jointly by clinicians and parent/patient partners and is structured to facilitate rapid quality improvement and learning^{3,4}. The survey summarized measure statements for 25 nutrition management guidelines^{5,6} and asked each respondent to rate level of agreement with the measure statement by a Likert scale. For example, to measure response by a dietitian assessment, we asked for agreement with the statement “if the patient has a visit with inadequate weight gain, we will measure if the patient has a dietitian assessment recorded at that visit or the subsequent visit.” We received responses from 15 physicians representing 12 programs. Twelve of 15 (80%) respondents were aware of the guideline definitions for expected average daily weight gain for infants. All measure statements had majority agreement from a range of 73% to 100%. Free text comments from respondents were also used to determine clarity of the measure statements. The study team used this input to refine the final practice definitions for the guidelines used in the analysis.

References

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eTable 1. Patients with at least 1 inadequate weight visit 0-24 months old

Characteristic^a	Total
Female	100 (46.3%)
Race/Ethnicity	
Black	12 (5.6%)
Caucasian	188 (87%)
Hispanic	11 (5.1%)
Other	5 (2.3%)
Genotype (n=215)	
Homozygous F508del	123 (57.2%)
Heterozygous F508del	79 (36.7%)
Other	13 (6.0%)
Insurance status (n=214)	
State or Federal Funded (e.g. Medicaid, Medicare, Indian Health Service)	74 (34.6%)
Private (e.g. commercial, HMO)	130 (60.7%)
Military (e.g. Tricare)	9 (4.2%)
Other	1 (0.5%)
Pancreatic insufficiency (n=211)	197 (93.4%)
History of meconium ileus	26 (11.5%)
Median age at diagnosis ^b , days (IQR)	8 (2-18)
Median birth weight, kg (IQR)	3.2 (2.9-3.5)
Ever Pseudomonas aeruginosa positive by 24 months old	102 (47.2%)
Median number of total visits per patient in 24 months (IQR)	13 (12-15)

^aN=216 unless otherwise noted

^bearliest date from sweat test or genetic testing date

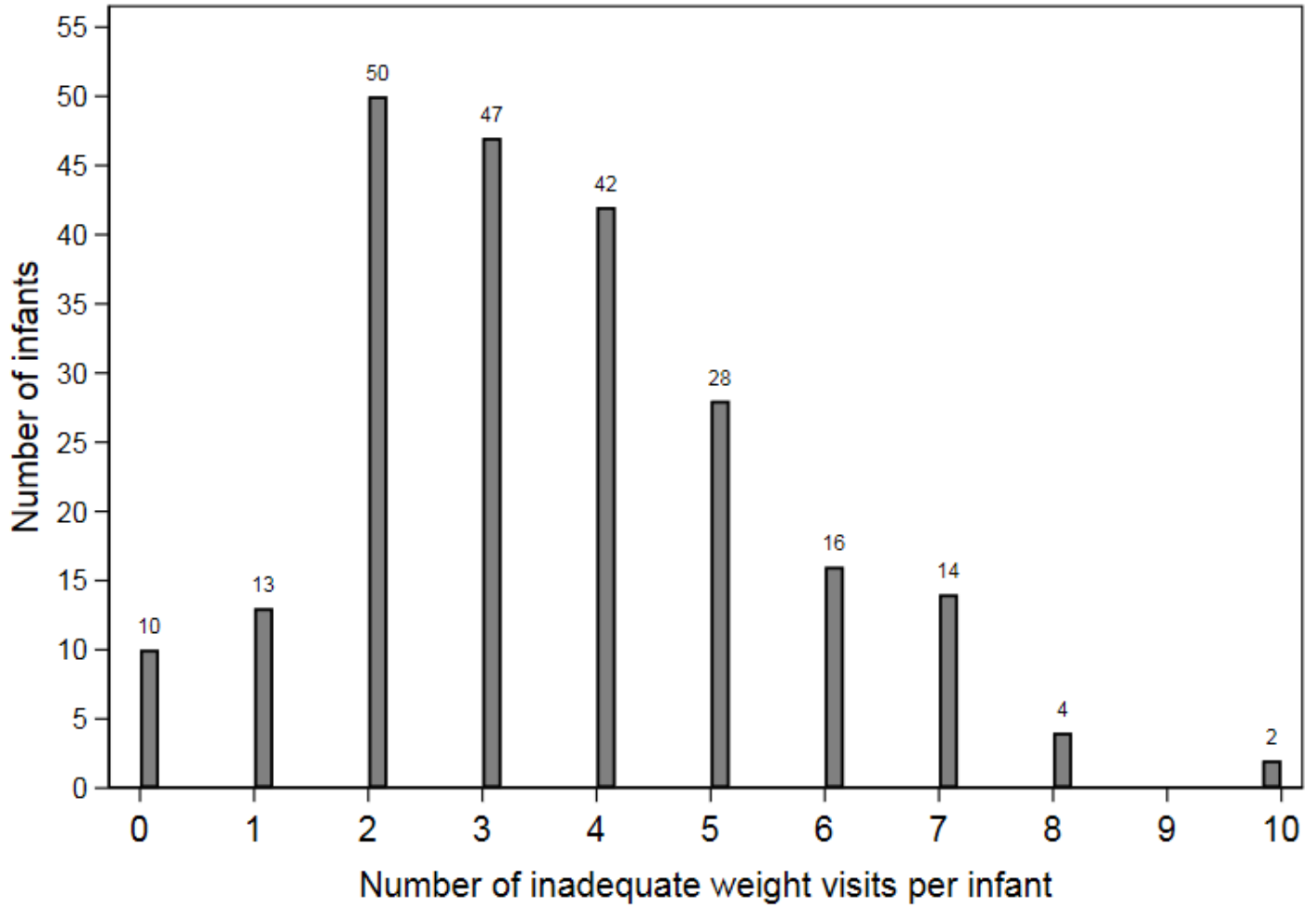
eTable 2. Patient Characteristics by Performance Group for Dietitian Assessments

Characteristic	Low RD sites	High RD sites	Mid RD sites (within limits)	p-value
Number of infants	34	39	153	0.28
Female	20 (58.8%)	16 (41.0%)	70 (48.8%)	
Race/Ethnicity				0.41
Black	4 (11.8%)	2 (5.1%)	6 (3.9%)	
Caucasian	28 (82.4%)	37 (94.9%)	132 (86.8%)	
Hispanic	2 (5.9%)	0 (0.0%)	9 (5.9%)	
Other	0 (0.0%)	0 (0.0%)	5 (2.6%)	
Genotype				0.22
Homozygous F508del	19 (55.9%)	23 (59.0%)	83 (54.6%)	
Heterozygous F508del	10 (29.4%)	14 (35.9%)	62 (40.8%)	
Other	5 (14.7%)	2 (5.1%)	7 (4.6%)	
Insurance status				0.75
State or Federal Funded (e.g. Medicaid, Medicare, Indian Health Service)	12 (36.4%)	17 (43.6%)	49 (32.0%)	
Private (e.g. commercial, HMO)	21 (63.6%)	20 (51.3%)	94 (61.8%)	
Military (e.g. Tricare)	0 (0.0%)	2 (5.1%)	7 (4.6%)	
Other	0 (0.0%)	0 (0.0%)	2 (1.3%)	
Pancreatic insufficiency ^a	33 (97.1%)	34 (89.5%)	137 (92.6%)	0.46
History of meconium ileus	7 (20.6%)	8 (20.5%)	11 (7.2%)	0.013
Median age at diagnosis ^b , days (IQR)	16.4 (0-29.2)	18.3 (11-25.6)	11.0 (3.7-20.1)	0.020
Mean birth weight, kg (SD)	3.2 (0.4)	3.3 (0.5)	3.2 (0.4)	0.57
Ever Pseudomonas aeruginosa positive by 24 months old	1 (2.9%)	1 (2.6%)	7 (4.6%)	0.75
Median number of clinic visits in 24 months (IQR)	11 (10-12)	13 (11-14)	11 (10-13)	0.006

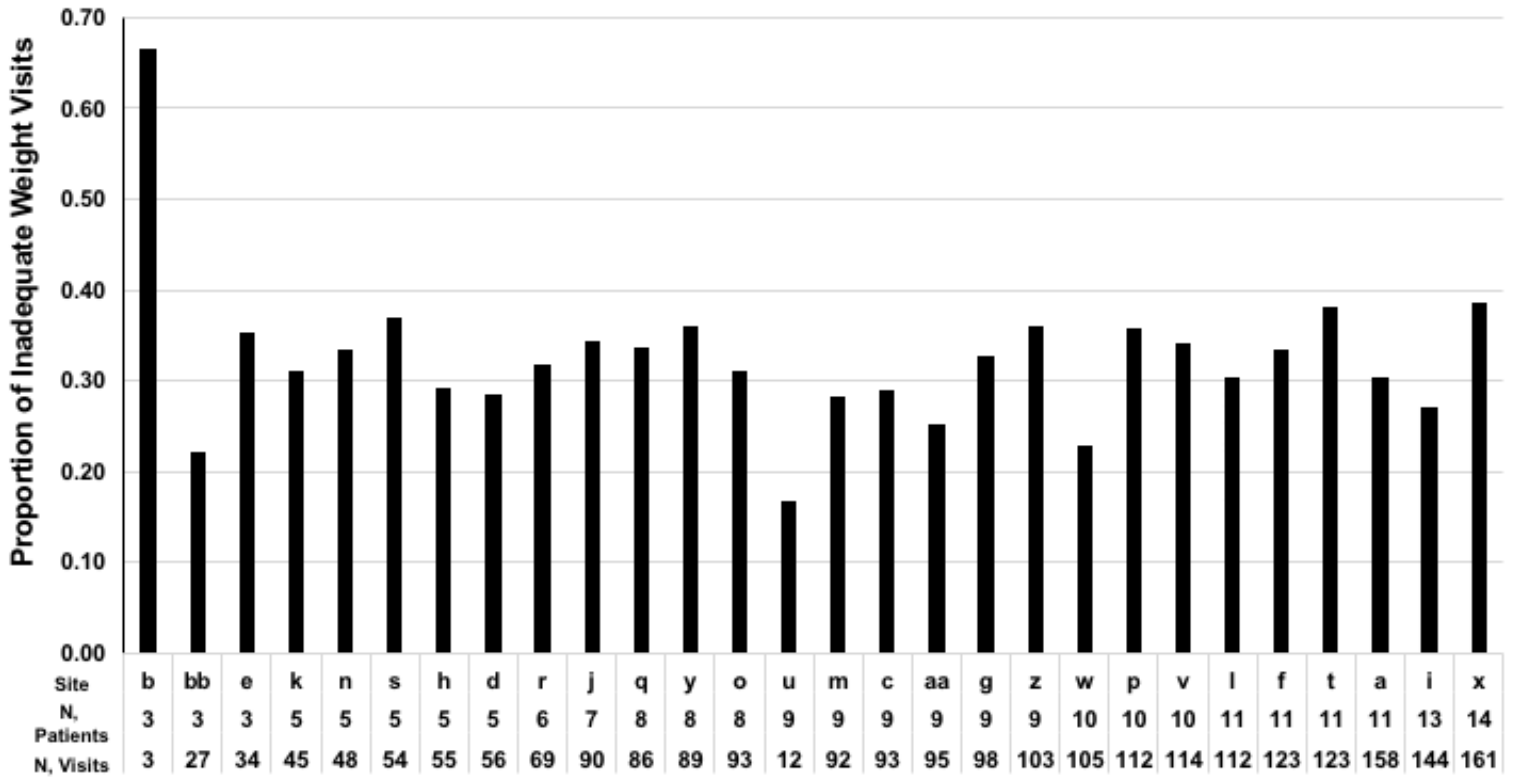
^aFecal elastase <=200 ug/g or 2 PI-causing CFTR mutations

^bearliest of sweat test or genetic testing date

eFigure 1. Distribution of inadequate weight gain visits per infant ≤ 24 months old



eFigure 2. Proportion of inadequate weight gain visits distributed across sites



eFigure 3. Distribution of WHO Weight-For-Age Z Score at 24-27 months old across Centers

