Case-Mix Adjustment in Cystic Fibrosis
A Model for Predicting Survival Based on Patient and Disease Characteristics Present at Diagnosis

G O'Connor, H Quinton, R Kahn, P Robichaud, J Maddock, T Lever, J Brooks

for the Northern New England Cystic Fibrosis Consortium

Supported by the Cystic Fibrosis Foundation

The Northern New England Cystic Fibrosis Consortium

The NNECFC is a regional, voluntary consortium of more than 70 clinicians and researchers from the CF care centers in Maine, New Hampshire and Vermont. The mission of the group is to improve CF care and patient outcomes.

Goal

Develop a case-mix adjustment model to predict survival for CF patients . A case-mix adjustment model contains patient and disease characteristics which are not a consequence of treatment but which are associated with survival.

Methods

The initial record from 27,565 patients in the US CFF National Patient Registry diagnosed before age 18 was used to develop a multivariate case-mix adjustment model.

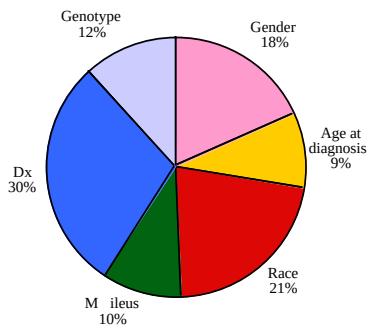
Cox proportional hazards regression was used to calculate adjusted hazard ratios (HR), 95% confidence intervals (CI), and tests of statistical significance.

The area under the relative operating characteristic curve (ROC) was used as a measure of discrimination of the model.

Predictors of decreased survival

Variable	HR	CI	P value
Female	1.36	1.28-1.44	<0.001
Dx at ≤ 6 mos. old	1.18	1.11-1.26	<0.001
Hispanic vs. White	1.89	1.58-2.25	<0.001
African-American vs. White	1.44	1.22-1.70	<0.001
Diagnosis suggested by:			
When compared to asymptomatic patients (diagnosed by genetic testing)			
Meconium ileus	1.67	1.38-2.02	<0.001
GI symptoms	1.32	1.11-1.56	<0.001
Respiratory symptoms	1.52	1.28-1.80	<0.001
Resp+GI symptoms	1.56	1.33-1.83	<0.001
Other symptoms	1.22	1.03-1.45	.020
Genotype when compared to ΔF508 heterozygotes			
ΔF508 homozygotes	1.29	1.12-1.48	<0.001
Neither mutation ΔF508	1.31	1.06-1.60	<0.001

Percent of predicted risk associated with patient and disease characteristics



Results

There were 390,855 patient years of follow-up and a total of 4,169 deaths.

The regression model was statistically significant
(X^2[df 15]=1939, p<0.0001

The ROC area was 0.73, indicating a moderately good ability to discriminate survival time.

Statistically significant predictors of decreased survival included gender, age at diagnosis, race/ethnicity, meconium ileus, criteria for CF diagnosis, and genotype (summarized in graph).

Conclusions

- This case-mix adjustment model uses patient and disease characteristics available at diagnosis to predict survival and has relatively good performance characteristics.
- Case-mix adjustment would allow benchmarking of clinical outcomes. This model is a step toward consensus on case-mix adjustment in cystic fibrosis.