

Predicting Survival Based upon Attained Age in Patients with Cystic Fibrosis

M Harder, H Quinton, T Lever, J Maddock, M Detzer, J Zuckerman, G O'Connor
for the NNECFC supported by the Cystic Fibrosis Foundation

The Northern New England Cystic Fibrosis Consortium



The NNECFC is a regional, voluntary consortium of more than 80 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

- To predict survival up to 10 years in cystic fibrosis patients, based on: attained age; gender; clinical findings.

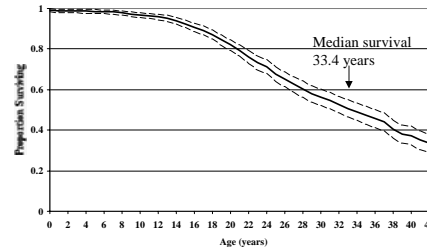
Methods

- Data set: all 35,510 patients with one or more annual records in the US CF Patient Registry from 1982 to 2001.
- Patients followed until death or censored at loss to follow up.
- Patient and disease characteristics included: sex, *P. aeruginosa*; FEV₁; body weight; CF-related insulin dependent diabetes (CFRD).
- Generalized gamma model was used to predict survival.

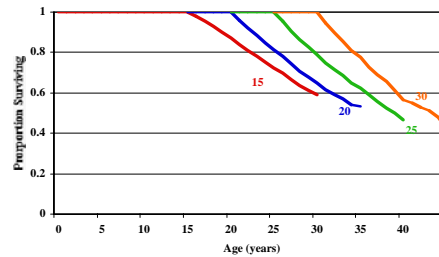
Results

- The fit of the gamma model was checked by comparing the parametric survival curves from the model to the Kaplan Meier survival curves for each covariate, separately and for some combinations of covariates for patients aged 15-30 years.
- The generalized gamma model with covariates sex, FEV₁ (<40%, 40 to 70%, >70%), CFRD, *P. aeruginosa*, and body weight (<10th percentile, 10th to 25th percentile, >25th percentile), was used to predict median survival beyond an attained age of 15 years.

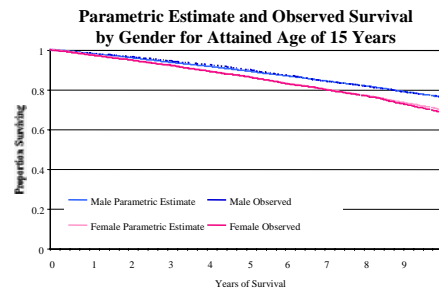
CFF Registry Actuarial Survival Prediction Curve for 2001
(with 95% confidence bounds)
(n=22,732 409 Deaths)



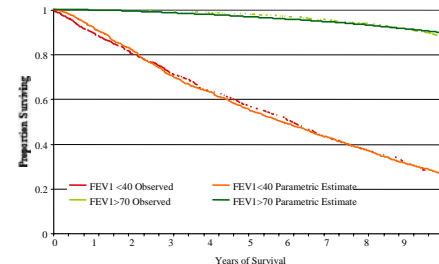
Observed 15 Year Survival Beyond the Attained Age
CFF Patient Registry 1982-2001



Goodness of Fit for Model
(n=11,370 2,347 Deaths)



Parametric Estimate and Observed Survival
Comparing FEV₁ <40% with FEV₁ >70%
for Attained Age of 15 Years



Predicted median survival times for an attained age of 15 years

Females FEV₁ % predicted

CDC Weight Percentile	<40%		40 to 70%		>70%	
	PA-	PA+	PA-	PA+	PA-	PA+
<10	CFRD - 5.0	3.7	12	8.6*	>20	>20
	CFRD + 2.9	2.1	6.9	5.0	16	12
10 to 25	CFRD - 8.0	5.8	19	14	>20	>20
	CFRD + 4.6	3.4	11	7.9	>20	19
>25	CFRD - 11	8.0	>20	19	>20	>20
	CFRD + 6.4	4.7	15	11	>20	>20

Males FEV₁ % predicted

CDC Weight Percentile	<40%		40 to 70%		>70%	
	PA-	PA+	PA-	PA+	PA-	PA+
<10	CFRD - 7.2	5.2	17	12	>20	>20
	CFRD + 4.2	3.1	9.9	7.2	>20	17
10 to 25	CFRD - 11	8.3	>20	19	>20	>20
	CFRD + 6.6	4.8	16	11	>20	>20
>25	CFRD - 16	11	>20	>20	>20	>20
	CFRD + 9.2	6.7	>20	16	>20	>20

***Example:** A female patient 15 years old with an FEV₁ of 60%, body weight under the 10th percentile, no diabetes, and positive *P. aeruginosa*, would have a predicted survival of 8.6 years (CI 95% = 8.0, 9.2) beyond her current age of 15.

Conclusions

- The generalized gamma model fits the data well for up to 10 years of survival beyond attained age based on the fitted survival curves.
- Median survival can be predicted for individual patients from a parametric model.
- More fine tuning in predicting survival is possible by adding other predictive covariates such as household income, liver disease, etc.