

**Table 14.2.6.1 Summary of Pulmonary Functions - by timepoint - FAS**

Parameter Timepoint	Actual value	Absolute change from baseline	% Change from baseline	t-Test results			
				Change from baseline	t-value (df)	p-value	95% CI
FVC (L)							
Baseline							
n	29						
Mean (SD)	2.651(1.083)						
Median	2.760						
Min; Max	0.87; 5.28						
Month 6							
n	22	20	20	Absolute change	2.749 (19)	0.013	(0.056,0.417)
Mean (SD)	2.714(1.055)	0.237(0.385)	11.36(21.57)				
Median	2.890	0.220	11.22	% Change	2.356 (19)	0.029	(1.270,21.46)
Min; Max	0.67; 4.36	-0.39; 1.20	-32.2;56.07				
Month 12							
n	30	28	28	Absolute change	4.876 (27)	<.001	(0.226,0.555)
Mean (SD)	3.102(1.170)	0.391(0.424)	16.33(19.56)				
Median	3.205	0.380	12.09	% Change	4.419 (27)	<.001	(8.748,23.91)
Min; Max	0.78; 5.80	-0.25; 1.38	-15.7;57.02				
Month 18							
n	8	8	8	Absolute change	2.445 ( 7)	0.044	(0.014,0.809)
Mean (SD)	2.688(1.052)	0.411(0.476)	17.14(23.95)				
Median	3.040	0.530	16.48	% Change	2.024 ( 7)	0.083	(-2.88,37.17)
Min; Max	1.21; 3.99	-0.53; 0.99	-30.5;46.26				
Month 24							
n	8	7	7	Absolute change	2.831 ( 6)	0.030	(0.117,1.614)
Mean (SD)	3.845(0.911)	0.866(0.809)	37.37(34.89)				
Median	3.715	0.970	41.25	% Change	2.833 ( 6)	0.030	(5.097,69.63)
Min; Max	2.21; 4.96	-0.08; 2.05	-2.09;79.77				

Note 1: FAS=Full Analysis Set. n=Number of subjects with data.

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Parameter	Timepoint	Actual value	Absolute change from baseline	% Change from baseline	t-Test results		
					Change from baseline	t-value (df)	p-value
FVC (L)							
Month 36							
n	6	6	6	Absolute change	3.354 ( 5)	0.020	(0.087,0.657)
Mean (SD)	2.353(0.897)	0.372(0.271)	25.24(25.02)				
Median	2.260	0.430	20.11	% Change	2.471 ( 5)	0.056	(-1.02,51.49)
Min; Max	1.20; 3.71	0.04; 0.69	1.84;68.32				
Month 48							
n	7	7	7	Absolute change	4.048 ( 6)	0.007	(0.433,1.756)
Mean (SD)	3.447(0.733)	1.094(0.715)	56.75(49.00)				
Median	3.580	0.820	54.55	% Change	3.064 ( 6)	0.022	(11.43,102.1)
Min; Max	1.87; 4.06	0.42; 2.48	13.21;157.0				
Last Observation							
n	31	29	29	Absolute change	4.417 (28)	<.001	(0.310,0.845)
Mean (SD)	3.233(1.153)	0.578(0.704)	28.38(37.79)				
Median	3.390	0.520	16.28	% Change	4.044 (28)	<.001	(14.00,42.76)
Min; Max	1.12; 5.80	-0.53; 2.48	-30.5;157.0				

Note 1: FAS=Full Analysis Set. n=Number of subjects with data.