

Listing 16.2.9.1 (Page 1 of 7)
 Demographics Characteristics and Baseline Characteristics
 (Matched Population for Piece-wise Regression Analysis on FVC)

Study: 019

| Subject ID | Baseline Age (yrs) | Sex | Race | Baseline FVC | Baseline %-Predicted FVC | Baseline %-Predicted FEV1 | Baseline PEF | Baseline EK Score |
|------------|--------------------|-----|-----------------|--------------|--------------------------|---------------------------|--------------|-------------------|
| 001013 | 17 | M | White/Caucasian | 2.56 | 98.62 | 95 | 4.49 | . |
| 003003 | 16 | M | Asian | 2.26 | 83.21 | 78.42 | 4.39 | . |
| 011001 | 16 | M | White/Caucasian | 1.52 | . | . | 2.3 | . |
| 011002 | 14 | M | White/Caucasian | 2.57 | 110.34 | 101.39 | 5 | 7 |
| 011003 | 17 | M | White/Caucasian | 2.73 | . | . | 5.69 | . |
| 011004 | 15 | M | White/Caucasian | 2.4 | 99 | 91.97 | 4.79 | 1 |
| 011005 | 13 | M | White/Caucasian | 2.22 | 94.97 | 87.78 | 4 | 9 |
| 011006 | 13 | M | White/Caucasian | 2.15 | 95.62 | 88.03 | 4 | 0 |
| 012001 | 15 | M | White/Caucasian | 2.14 | 76.32 | 68.37 | . | 9 |
| 012005 | 15 | M | White/Caucasian | 1.48 | 53.31 | 55.3 | 1.16 | . |
| 021001 | 14 | M | White/Caucasian | 2.09 | 69.63 | 66.12 | 4 | 4 |
| 021004 | 16 | M | White/Caucasian | 0.85 | 30.25 | 28.99 | 2.14 | . |
| 021005 | 17 | M | White/Caucasian | 2.24 | 72.79 | 72.91 | 5.36 | . |
| 031001 | 21 | M | White/Caucasian | 1.16 | 32.57 | 28.67 | 3 | 12 |
| 031002 | 15 | M | White/Caucasian | 1.6 | 71.83 | 64.35 | 3.3 | . |
| 031003 | 11 | M | White/Caucasian | 1.31 | 58.05 | 58.3 | 2.91 | 9 |
| 034001 | 13 | M | White/Caucasian | 1.29 | 61.9 | 64.58 | 3 | . |
| 034002 | 14 | M | Asian | 1.83 | 71.28 | 71.84 | 4 | . |
| 041004 | 13 | M | White/Caucasian | 1.83 | 72.36 | 69.49 | 3 | 3 |
| 041005 | 12 | M | White/Caucasian | 1.61 | 63.57 | 63.25 | 2 | 8 |
| 042001 | 11 | M | White/Caucasian | 1.71 | 78.07 | 63.67 | 2 | 8 |
| 042002 | 12 | M | White/Caucasian | 2.42 | 82.03 | 82.45 | 5 | 0 |
| 043001 | 12 | M | White/Caucasian | 2.01 | 80.06 | 75.76 | 4 | 11 |
| 043002 | 14 | M | Asian | 2.54 | 80.28 | 63.53 | 4 | 4 |
| 051001 | 20 | M | White/Caucasian | 1.16 | 46.17 | 33.56 | 1.3 | 11 |

Listing 16.2.9.1 (Page 2 of 7)
 Demographics Characteristics and Baseline Characteristics
 (Matched Population for Piece-wise Regression Analysis on FVC)

Study: 019

| Subject ID | Baseline Age (yrs) | Sex | Race | Baseline FVC | Baseline %-Predicted FVC | Baseline %-Predicted FEV1 | Baseline PEF | Baseline EK Score |
|------------|--------------------|-----|-----------------|--------------|--------------------------|---------------------------|--------------|-------------------|
| 051003 | 15 | M | White/Caucasian | 2.44 | 62.18 | 62.01 | 4 | 7 |
| 052006 | 11 | M | White/Caucasian | 2.24 | 86.27 | 80.54 | 3.49 | 5 |
| 061001 | 12 | M | White/Caucasian | 1.93 | 84.89 | 81.41 | 218 | 4 |
| 061002 | 12 | M | White/Caucasian | 1.94 | 77.25 | 80.24 | 3 | . |
| 062001 | 14 | M | White/Caucasian | 2.52 | 68.62 | 73.51 | 3 | 8 |
| 062002 | 15 | M | White/Caucasian | 1.94 | . | . | 3.16 | . |
| 062004 | 15 | M | White/Caucasian | 2.08 | 62.66 | 65.9 | 4.25 | . |
| 063001 | 15 | M | White/Caucasian | 1.97 | 70.29 | 70.58 | 3.35 | . |
| 063002 | 13 | M | Other | 2.48 | 73.44 | 73.78 | 4 | 7 |
| 063005 | 12 | M | White/Caucasian | 1.89 | 80.75 | 77.9 | 4.72 | . |
| 063006 | 9 | M | White/Caucasian | 1.53 | 74.82 | 68.25 | 3 | 6 |
| 071001 | 12 | M | White/Caucasian | 1.67 | 66.15 | 67.24 | 4 | 10 |
| 071002 | 13 | M | White/Caucasian | 1.81 | 75.76 | 74.38 | 4.06 | . |
| 081001 | 14 | M | White/Caucasian | 1.66 | 77.44 | 76.18 | 3.75 | . |
| 081002 | 10 | M | White/Caucasian | 1.79 | 83.87 | 82.56 | 7.08 | 2 |
| 082003 | 14 | M | White/Caucasian | 2.38 | 103.83 | 98.84 | 3 | 7 |
| 082004 | 14 | M | White/Caucasian | 2.45 | 83.99 | 64.77 | 3 | 14 |
| 091001 | 12 | M | White/Caucasian | 1.83 | 75.33 | 72.9 | 3.04 | . |
| 091002 | 18 | M | White/Caucasian | 2.58 | 70.74 | 61.29 | 5 | 3 |
| 091003 | 11 | M | White/Caucasian | 1.12 | 53.71 | 47.6 | 1.45 | . |
| 091004 | 15 | M | White/Caucasian | 1.09 | 46.6 | 42.1 | 3 | 6 |
| 092001 | 13 | M | White/Caucasian | 0.97 | . | . | 2.72 | 1 |
| 092002 | 15 | M | White/Caucasian | 1.45 | 59.9 | 50.36 | 1.66 | 7 |
| 092003 | 14 | M | Other | 1.46 | 51.61 | 50.45 | 3.46 | 4 |
| 101002 | 15 | M | White/Caucasian | 1.25 | . | . | 3.79 | . |

Listing 16.2.9.1 (Page 3 of 7)
 Demographics Characteristics and Baseline Characteristics
 (Matched Population for Piece-wise Regression Analysis on FVC)

Study: 019

| Subject ID | Baseline Age (yrs) | Sex | Race | Baseline FVC | Baseline %-Predicted FVC | Baseline %-Predicted FEV1 | Baseline PEF | Baseline EK Score |
|------------|--------------------|-----|-----------------|--------------|--------------------------|---------------------------|--------------|-------------------|
| 101005 | 16 | M | White/Caucasian | 1.34 | 45.54 | 42.91 | 2.21 | . |
| 101006 | 17 | M | White/Caucasian | 2.65 | . | . | 3.4 | . |
| 101007 | 13 | M | White/Caucasian | 1.71 | . | . | . | . |
| 101008 | 14 | M | White/Caucasian | 1.42 | . | . | 2.92 | 10 |
| 102001 | 15 | M | Asian | 2.72 | 86.61 | 87.99 | 4.08 | . |
| 102002 | 13 | M | White/Caucasian | 1.49 | . | . | 3.86 | 5 |
| 103001 | 14 | M | White/Caucasian | 1.88 | . | . | 3.48 | 12 |
| 103003 | 16 | M | White/Caucasian | 2.79 | 87.42 | 74.01 | 4.14 | 12 |
| 103004 | 16 | M | White/Caucasian | 1.1 | . | . | 3.32 | . |
| 103006 | 15 | M | White/Caucasian | 3.62 | . | . | 5.87 | . |

Listing 16.2.9.1 (Page 4 of 7)
 Demographics Characteristics and Baseline Characteristics
 (Matched Population for Piece-wise Regression Analysis on FVC)

Study: CINRG

| Subject ID | Baseline Age (yrs) | Sex | Race | Baseline FVC | Baseline %-Predicted FVC | Baseline %-Predicted FEV1 | Baseline PEF | Baseline EK Score |
|------------|--------------------|-----|-----------------|--------------|--------------------------|---------------------------|--------------|-------------------|
| 042 | 20 | M | White/Caucasian | 2.12 | 42 | 47 | 4.82 | . |
| 049 | 18 | M | White/Caucasian | 2.15 | 56 | 57 | 4.93 | . |
| 051 | 18 | M | White/Caucasian | 1.87 | 44 | 36 | 4.99 | . |
| 055 | 19 | M | White/Caucasian | 1.31 | 29 | 33 | 2.99 | . |
| 060 | 18 | M | White/Caucasian | 1.73 | 39 | 40 | 3.43 | . |
| 066 | 17 | M | White/Caucasian | 0.73 | 15 | 17 | 1.68 | . |
| 067 | 17 | M | White/Caucasian | 0.85 | 19 | 21 | 1.4 | . |
| 068 | 17 | M | White/Caucasian | 1.48 | 32 | 32 | 2.04 | . |
| 072 | 17 | M | White/Caucasian | 1.09 | 25 | 25 | 1.91 | . |
| 078 | 15 | M | White/Caucasian | 3.86 | 83 | 92 | 6.77 | . |
| 079 | 15 | M | Asian | 2.38 | 69 | 67 | 4.44 | . |
| 088 | 16 | M | White/Caucasian | 1.96 | 55 | 61 | 4.1 | . |
| 089 | 15 | M | White/Caucasian | 4.84 | 101 | 103 | 7.33 | . |
| 090 | 15 | M | White/Caucasian | 0.84 | 30 | 28 | 2.99 | . |
| 094 | 15 | M | White/Caucasian | 2.35 | 91 | 83 | 4.65 | . |
| 096 | 14 | M | White/Caucasian | 2.18 | 78 | 76 | 4.44 | . |
| 114 | 14 | M | White/Caucasian | 1.55 | 60 | 56 | 3.26 | . |
| 120 | 13 | M | White/Caucasian | 2.73 | 94 | 101 | 4.2 | . |
| 121 | 13 | M | Other | 2.33 | 69 | 72 | 3.68 | . |
| 122 | 13 | M | Asian | 1.48 | 68 | 56 | 2.12 | . |
| 123 | 13 | M | White/Caucasian | 1.72 | 69 | 55 | 3.1 | . |
| 126 | 14 | M | White/Caucasian | 1.42 | 53 | 56 | 3.57 | . |
| 128 | 13 | M | White/Caucasian | 1.9 | 73 | 73 | 3.78 | . |
| 131 | 12 | M | White/Caucasian | 2.31 | 66 | 75 | 4.96 | . |
| 133 | 13 | M | White/Caucasian | 2.17 | 64 | 65 | 3.12 | . |

Listing 16.2.9.1 (Page 5 of 7)
 Demographics Characteristics and Baseline Characteristics
 (Matched Population for Piece-wise Regression Analysis on FVC)

Study: CINRG

| Subject ID | Baseline Age (yrs) | Sex | Race | Baseline FVC | Baseline %-Predicted FVC | Baseline %-Predicted FEV1 | Baseline PEF | Baseline EK Score |
|------------|--------------------|-----|-----------------|--------------|--------------------------|---------------------------|--------------|-------------------|
| 134 | 11 | M | White/Caucasian | 1.84 | 73 | 74 | 2.78 | . |
| 135 | 12 | M | White/Caucasian | 2.75 | 86 | 82 | 4.53 | . |
| 136 | 12 | M | Asian | 1.43 | 53 | 54 | 3.69 | . |
| 140 | 12 | M | White/Caucasian | 1.03 | 39 | 41 | 3.04 | . |
| 144 | 12 | M | White/Caucasian | 1.62 | 94 | 87 | 4.31 | . |
| 147 | 12 | M | White/Caucasian | 2.19 | 73 | 65 | 3.99 | . |
| 148 | 11 | M | White/Caucasian | 1.53 | 86 | 86 | 3.41 | . |
| 152 | 11 | M | White/Caucasian | 1.24 | 61 | 64 | 2.78 | . |
| 157 | 12 | M | White/Caucasian | 1.64 | 66 | 67 | 3.05 | . |
| 159 | 12 | M | White/Caucasian | 1.86 | 74 | 78 | 4.02 | . |
| 160 | 11 | M | White/Caucasian | 2.14 | 75 | 66 | 3.24 | . |
| 161 | 11 | M | White/Caucasian | 2.39 | 106 | 82 | 2.38 | . |
| 169 | 11 | M | Other | 1.64 | 64 | 67 | 3.16 | . |
| 170 | 12 | M | White/Caucasian | 1.24 | 76 | 74 | 2.87 | . |
| 179 | 10 | M | White/Caucasian | 1.51 | 68 | 64 | 2.63 | . |
| 181 | 10 | M | White/Caucasian | 1.9 | 134 | 124 | 4.06 | . |
| 182 | 10 | M | White/Caucasian | 2.7 | 88 | 89 | 5.23 | . |
| 183 | 12 | M | Other | 1.7 | 53 | 59 | 2.68 | . |
| 184 | 10 | M | White/Caucasian | 1.6 | 87 | 80 | 2.56 | . |
| 187 | 10 | M | White/Caucasian | 1.6 | 63 | 73 | 3.05 | . |
| 189 | 10 | M | White/Caucasian | 1.43 | 82 | 78 | 2.46 | . |
| 190 | 9 | M | White/Caucasian | 1.12 | 60 | 52 | 2.55 | . |
| 191 | 10 | M | Other | 2.02 | 113 | 103 | 2.65 | . |
| 193 | 10 | M | White/Caucasian | 1.25 | 79 | 77 | 3.01 | . |
| 194 | 9 | M | White/Caucasian | 1.49 | 80 | 72 | 1.42 | . |

Listing 16.2.9.1 (Page 6 of 7)
 Demographics Characteristics and Baseline Characteristics
 (Matched Population for Piece-wise Regression Analysis on FVC)

Study: CINRG

| Subject ID | Baseline Age (yrs) | Sex | Race | Baseline FVC | Baseline %-Predicted FVC | Baseline %-Predicted FEV1 | Baseline PEF | Baseline EK Score |
|------------|--------------------|-----|------------------|--------------|--------------------------|---------------------------|--------------|-------------------|
| 195 | 11 | M | Other | 2.21 | 98 | 96 | 3.08 | . |
| 198 | 11 | M | White/Caucasian | 2.01 | 64 | 66 | 3.95 | . |
| 199 | 9 | M | White/Caucasian | 1.63 | 93 | 77 | 1.59 | . |
| 202 | 8 | M | White/Caucasian | 1.54 | 84 | 87 | 2.25 | . |
| 203 | 8 | M | White/Caucasian | 1.77 | 121 | 123 | 3.24 | . |
| 204 | 9 | M | White/Caucasian | 1.35 | 68 | 70 | 2.93 | . |
| 205 | 9 | M | White/Caucasian | 2.33 | 106 | 102 | 3.02 | . |
| 206 | 10 | M | White/Caucasian | 2.22 | 140 | 114 | 3.96 | . |
| 207 | 8 | M | White/Caucasian | 1.15 | 83 | 78 | 1.19 | . |
| 210 | 8 | M | White/Caucasian | 1.69 | 95 | 89 | 1.93 | . |
| 217 | 9 | M | White/Caucasian | 1.63 | 76 | 65 | 2.16 | . |
| 219 | 7 | M | Pacific Islander | 1.77 | 113 | 105 | 2.88 | . |
| 222 | 8 | M | White/Caucasian | 1.95 | 96 | 98 | 2.05 | . |
| 225 | 8 | M | Asian | 1.15 | 59 | 70 | 2.01 | . |
| 227 | 9 | M | Asian | 1.11 | 86 | 76 | 1.84 | . |
| 228 | 9 | M | White/Caucasian | 1.88 | 93 | 105 | 3.49 | . |
| 231 | 10 | M | White/Caucasian | 1.43 | 82 | 84 | 2.99 | . |
| 233 | 9 | M | White/Caucasian | 1.78 | 70 | 79 | 3.41 | . |
| 234 | 9 | M | White/Caucasian | 2.28 | 98 | 105 | 3.67 | . |
| 240 | 9 | M | Asian | 0.84 | 50 | 54 | 1.3 | . |
| 241 | 8 | M | White/Caucasian | 1.54 | 93 | 105 | 3.48 | . |
| 243 | 10 | M | Asian | 1.87 | 77 | 77 | 2.65 | . |
| 252 | 7 | M | White/Caucasian | 1.1 | 73 | 87 | 2.3 | . |
| 258 | 7 | M | White/Caucasian | 1.32 | 111 | 130 | 2.63 | . |
| 261 | 7 | M | White/Caucasian | 1.59 | 94 | 114 | 4.01 | . |

Listing 16.2.9.1 (Page 7 of 7)
 Demographics Characteristics and Baseline Characteristics
 (Matched Population for Piece-wise Regression Analysis on FVC)

Study: CINRG

| Subject ID | Baseline Age (yrs) | Sex | Race | Baseline FVC | Baseline %-Predicted FVC | Baseline %-Predicted FEV1 | Baseline PEF | Baseline EK Score |
|------------|--------------------|-----|-----------------|--------------|--------------------------|---------------------------|--------------|-------------------|
| 266 | 7 | M | White/Caucasian | 1.36 | 87 | 105 | 2.67 | . |
| 272 | 6 | M | White/Caucasian | 1.36 | 95 | 110 | 3.44 | . |
| 274 | 7 | M | Other | 1.06 | 74 | 73 | 1.97 | . |
| 275 | 7 | M | Other | 1.49 | 106 | 106 | 1.75 | . |
| 283 | 6 | M | Other | 1.17 | 72 | 83 | 2.23 | . |
| 284 | 6 | M | White/Caucasian | 1.49 | 97 | 100 | 2.11 | . |
| 285 | 6 | M | White/Caucasian | 1.07 | 90 | 93 | 2.34 | . |
| 293 | 6 | M | White/Caucasian | 1.41 | 92 | 116 | 2.75 | . |
| 297 | 6 | M | White/Caucasian | 1.18 | 96 | 115 | 2.06 | . |
| 300 | 6 | M | White/Caucasian | 0.89 | 55 | 51 | 1.89 | . |
| 316 | 7 | M | White/Caucasian | 1.43 | 94 | 107 | 2.64 | . |
| 317 | 6 | M | White/Caucasian | 1.56 | 95 | 107 | 2.78 | . |
| 321 | 7 | M | White/Caucasian | 1.2 | 100 | 113 | 2.1 | . |
| 322 | 6 | M | White/Caucasian | 1.43 | 98 | 122 | 2.7 | . |
| 325 | 7 | M | White/Caucasian | 1.31 | 91 | 97 | 2.76 | . |
| 342 | 7 | M | White/Caucasian | 1.63 | 108 | 121 | 3.32 | . |

Listing 16.2.9.2 (Page 1 of 5)
 Demographics Characteristics and Baseline Characteristics
 (Matched Population for Age at FVC < 1 Liter Analysis)

Study: 019

| Subject ID | Baseline Age (yrs) | Race | Steroid Use | Baseline corticosteroid type | Cumul. BL Steroid Duration | Baseline FVC (L) | Baseline %-predicted FVC | Baseline %-predicted FEV1 | Baseline PEF | Baseline EK Score |
|------------|--------------------|-----------------|-------------|------------------------------|----------------------------|------------------|--------------------------|---------------------------|--------------|-------------------|
| 011002 | 14.06 | White/Caucasian | Y | Prednisone/Prednisolone | >=12 month | 2.57 | 110.34 | 101.39 | 5 | 7 |
| 011004 | 15.49 | White/Caucasian | Y | Prednisone/Prednisolone | >=12 month | . | . | . | . | 1 |
| 011005 | 13.41 | White/Caucasian | Y | Prednisone/Prednisolone | >=12 month | 2.22 | 94.97 | 87.78 | 4 | 9 |
| 011006 | 13.07 | White/Caucasian | Y | Prednisone/Prednisolone | <12 month | 2.15 | 95.62 | 88.03 | 4 | 0 |
| 012001 | 15.73 | White/Caucasian | Y | Deflazacort | >=12 month | 2.14 | 76.38 | 76.56 | 4.83 | 9 |
| 021001 | 14.77 | White/Caucasian | Y | Deflazacort | >=12 month | 2.09 | 69.67 | 66.15 | 4 | 4 |
| 031003 | 11.78 | White/Caucasian | Y | Deflazacort | >=12 month | 1.31 | 58.05 | 58.3 | 2.91 | 9 |
| 041004 | 13.59 | White/Caucasian | Y | Prednisone/Prednisolone | >=12 month | 1.83 | 72.41 | 69.54 | 4 | 3 |
| 041005 | 12.69 | White/Caucasian | N | None | <12 month | 1.61 | 63.62 | 63.73 | 3 | 8 |
| 042001 | 11.03 | White/Caucasian | Y | Prednisone/Prednisolone | >=12 month | 1.71 | 78.26 | 63.83 | 2 | 8 |
| 042002 | 12.65 | White/Caucasian | Y | Prednisone/Prednisolone | >=12 month | 2.42 | 82.23 | 82.67 | 5 | 0 |
| 043001 | 12.29 | White/Caucasian | Y | Prednisone/Prednisolone | >=12 month | 2.01 | 80.13 | 76.67 | 4 | 11 |
| 043002 | 14.36 | Asian | Y | Prednisone/Prednisolone | >=12 month | 2.54 | 80.35 | 64.92 | 4 | 4 |
| 051003 | 15.87 | White/Caucasian | Y | Prednisone/Prednisolone | >=12 month | 2.44 | 62.18 | 62.01 | 4 | 7 |
| 052002 | 15.2 | White/Caucasian | N | None | <12 month | 1.99 | 63.17 | 56.34 | 3.38 | 11 |
| 052003 | 12.98 | White/Caucasian | N | None | <12 month | 2.86 | 103.63 | 104 | 6 | 11 |
| 052006 | 11.48 | White/Caucasian | Y | Prednisone/Prednisolone | >=12 month | 2.24 | 86.27 | 80.54 | 3.49 | 5 |
| 061001 | 12 | White/Caucasian | Y | Deflazacort | >=12 month | 1.93 | 84.89 | 81.41 | 218 | 4 |
| 062001 | 14.28 | White/Caucasian | Y | Deflazacort | >=12 month | 2.52 | 68.62 | 73.51 | 3 | 8 |
| 062003 | 13.6 | White/Caucasian | N | None | <12 month | 2.48 | 72.16 | 72.34 | 3 | 9 |
| 063002 | 13.21 | Other | Y | Deflazacort | >=12 month | 2.48 | 73.44 | 74.42 | 4 | 7 |
| 063006 | 9.41 | White/Caucasian | Y | Deflazacort | >=12 month | 1.53 | 74.82 | 72.41 | 3 | 6 |
| 071001 | 12.63 | White/Caucasian | Y | Prednisone/Prednisolone | >=12 month | 1.67 | 66.15 | 67.24 | 4 | 10 |
| 081002 | 10.22 | White/Caucasian | Y | Deflazacort | >=12 month | 1.79 | 83.87 | 82.56 | 7.08 | 2 |
| 082003 | 14.05 | White/Caucasian | Y | Prednisone/Prednisolone | >=12 month | 2.38 | 103.83 | 98.84 | 3 | 7 |

Listing 16.2.9.2 (Page 2 of 5)
 Demographics Characteristics and Baseline Characteristics
 (Matched Population for Age at FVC < 1 Liter Analysis)

Study: 019

| Subject ID | Baseline Age (yrs) | Race | Steroid Use | Baseline corticosteroid type | Cumul. BL Steroid Duration | Baseline FVC (L) | Baseline %-predicted FVC | Baseline %-predicted FEV1 | Baseline PEF | Baseline EK Score |
|------------|--------------------|-----------------|-------------|------------------------------|----------------------------|------------------|--------------------------|---------------------------|--------------|-------------------|
| 082004 | 14.33 | White/Caucasian | Y | Prednisone/Prednisolone | >=12 month | 2.45 | 83.99 | 64.77 | 3 | 14 |
| 091004 | 15.27 | White/Caucasian | Y | Deflazacort | >=12 month | 1.09 | 46.6 | 42.1 | 3 | 6 |
| 092002 | 15.73 | White/Caucasian | Y | Deflazacort | >=12 month | 1.45 | 59.9 | 50.36 | 1.66 | 7 |
| 092003 | 14.28 | Other | Y | Prednisone/Prednisolone | >=12 month | 1.46 | 51.61 | 50.45 | 3.46 | 4 |
| 101004 | 10.86 | White/Caucasian | N | None | <12 month | 1.52 | . | . | 3.4 | 14 |
| 101007 | 12.8 | White/Caucasian | Y | Deflazacort | >=12 month | . | . | . | . | . |
| 101008 | 14.28 | White/Caucasian | Y | Deflazacort | >=12 month | 1.42 | . | . | 3.13 | 10 |
| 102002 | 13.91 | White/Caucasian | N | None | >=12 month | 1.49 | . | . | 3.86 | 5 |
| 102003 | 12.33 | White/Caucasian | N | None | <12 month | 1.64 | 65.75 | 61.81 | 3.37 | 4 |
| 103001 | 14.98 | White/Caucasian | Y | Prednisone/Prednisolone | >=12 month | 1.88 | . | . | 3.48 | 12 |
| 103003 | 16.46 | White/Caucasian | Y | Deflazacort | >=12 month | 2.79 | 87.42 | 74.01 | 4.14 | 12 |
| 103007 | 13.6 | White/Caucasian | N | None | <12 month | 1.79 | 85.79 | 80.51 | 2.86 | 15 |

Listing 16.2.9.2 (Page 3 of 5)
 Demographics Characteristics and Baseline Characteristics
 (Matched Population for Age at FVC < 1 Liter Analysis)

Study: CINRG

| Subject ID | Baseline Age (yrs) | Race | Steroid Use | Baseline corticosteroid type | Cumul. BL Steroid Duration | Baseline FVC (L) | Baseline %-predicted FVC | Baseline %-predicted FEV1 | Baseline PEF | Baseline EK Score |
|------------|--------------------|------------------------|-------------|------------------------------|----------------------------|------------------|--------------------------|---------------------------|--------------|-------------------|
| 052 | 17.83 | White/Caucasian | N | None | >=12 month | 1.12 | 27 | 30 | 2.87 | . |
| 068 | 17.17 | White/Caucasian | N | None | >=12 month | 1.48 | 32 | 32 | 2.04 | . |
| 071 | 16.18 | White/Caucasian | N | None | <12 month | 1.43 | 31 | 35 | 3.21 | . |
| 072 | 17.88 | White/Caucasian | Y | Deflazacort | >=12 month | 1.09 | 25 | 25 | 1.91 | . |
| 074 | 17.8 | Other | N | None | <12 month | 1.44 | 29 | 33 | 4.14 | . |
| 077 | 16.93 | White/Caucasian | N | None | >=12 month | 1.99 | 43 | 42 | 3.35 | . |
| 079 | 15.62 | Asian | Y | Deflazacort | >=12 month | 2.38 | 69 | 67 | 4.44 | . |
| 081 | 15.7 | Other | N | None | <12 month | 1.96 | 54 | 59 | 4.33 | . |
| 082 | 16.26 | White/Caucasian | N | None | >=12 month | 1.56 | 35 | 35 | 2.54 | . |
| 084 | 15.8 | White/Caucasian | N | None | <12 month | 2.77 | 60 | 64 | 5.84 | . |
| 088 | 16.37 | White/Caucasian | Y | Deflazacort | >=12 month | 1.96 | 55 | 61 | 4.1 | . |
| 091 | 14.58 | Asian | N | None | <12 month | 2.97 | 61 | 70 | 5.88 | . |
| 093 | 15.61 | Other | N | None | <12 month | 1.17 | 28 | 28 | 2.1 | . |
| 095 | 15.25 | White/Caucasian | N | None | <12 month | 1.66 | 36 | 38 | 2.66 | . |
| 096 | 14.45 | White/Caucasian | Y | Deflazacort | >=12 month | 2.18 | 78 | 76 | 4.44 | . |
| 099 | 14.82 | Asian | Y | Deflazacort | >=12 month | 1.38 | 62 | 64 | 2.89 | . |
| 102 | 14.53 | Black/African American | N | None | <12 month | 1.75 | 52 | 45 | 4.14 | . |
| 106 | 13.67 | White/Caucasian | N | None | >=12 month | 1.59 | 43 | 46 | 3.31 | . |
| 109 | 13.38 | White/Caucasian | Y | Prednisone/Prednisolone | >=12 month | 2.76 | 78 | 70 | 3.83 | . |
| 111 | 15.25 | White/Caucasian | N | None | <12 month | 2.28 | 62 | 68 | 3.99 | . |
| 115 | 12.78 | Other | N | None | <12 month | 1.67 | 42 | 34 | 2.71 | . |
| 117 | 13.68 | White/Caucasian | N | None | <12 month | 2.67 | 63 | 70 | 5.55 | . |
| 118 | 12.68 | Other | Y | Prednisone/Prednisolone | >=12 month | 1.97 | 69 | 73 | 3.64 | . |
| 121 | 13.74 | Other | Y | Prednisone/Prednisolone | >=12 month | 2.33 | 69 | 72 | 3.68 | . |
| 123 | 13.34 | White/Caucasian | Y | Deflazacort | >=12 month | 1.72 | 69 | 55 | 3.1 | . |

Listing 16.2.9.2 (Page 4 of 5)
 Demographics Characteristics and Baseline Characteristics
 (Matched Population for Age at FVC < 1 Liter Analysis)

Study: CINRG

| Subject ID | Baseline Age (yrs) | Race | Steroid Use | Baseline corticosteroid type | Cumul. BL Steroid Duration | Baseline FVC (L) | Baseline %-predicted FVC | Baseline %-predicted FEV1 | Baseline PEF | Baseline EK Score |
|------------|--------------------|-----------------|-------------|------------------------------|----------------------------|------------------|--------------------------|---------------------------|--------------|-------------------|
| 125 | 12.78 | White/Caucasian | Y | Deflazacort | >=12 month | 1.43 | 52 | 54 | 3.5 | . |
| 126 | 14.33 | White/Caucasian | Y | Prednisone/Prednisolone | >=12 month | 1.42 | 53 | 56 | 3.57 | . |
| 132 | 12.91 | White/Caucasian | N | None | <12 month | 2.39 | 69 | 70 | 4.44 | . |
| 134 | 11.74 | White/Caucasian | Y | Deflazacort | >=12 month | 1.84 | 73 | 74 | 2.78 | . |
| 136 | 12.95 | Asian | Y | Deflazacort | >=12 month | 1.43 | 53 | 54 | 3.69 | . |
| 137 | 12.02 | Other | Y | Prednisone/Prednisolone | >=12 month | 1.97 | 77 | 81 | 3.59 | . |
| 138 | 14.11 | Asian | N | None | <12 month | 1.35 | 35 | 37 | 2.09 | . |
| 139 | 14.11 | Asian | N | None | >=12 month | 1.8 | 43 | 46 | 3.49 | . |
| 140 | 12.65 | White/Caucasian | Y | Prednisone/Prednisolone | >=12 month | 1.03 | 39 | 41 | 3.04 | . |
| 150 | 12.11 | White/Caucasian | Y | Prednisone/Prednisolone | >=12 month | 1.42 | 69 | 73 | 3.37 | . |
| 157 | 12.51 | White/Caucasian | Y | Prednisone/Prednisolone | >=12 month | 1.64 | 66 | 67 | 3.05 | . |
| 159 | 12.82 | White/Caucasian | Y | Deflazacort | >=12 month | 1.86 | 74 | 78 | 4.02 | . |
| 160 | 11.32 | White/Caucasian | N | None | <12 month | 2.14 | 75 | 66 | 3.24 | . |
| 162 | 11.36 | Asian | N | None | <12 month | 1.21 | 46 | 51 | 2.35 | . |
| 163 | 11.91 | Other | N | None | <12 month | 1.55 | 50 | 52 | 2.26 | . |
| 164 | 10.45 | Asian | N | None | >=12 month | 1.52 | 74 | 75 | 4.17 | . |
| 169 | 11.48 | Other | N | None | >=12 month | 1.64 | 64 | 67 | 3.16 | . |
| 170 | 12.17 | White/Caucasian | Y | Deflazacort | >=12 month | 1.24 | 76 | 74 | 2.87 | . |
| 174 | 11.68 | Asian | N | None | <12 month | 1.72 | 56 | 64 | 3.39 | . |
| 175 | 12.36 | Asian | Y | Prednisone/Prednisolone | >=12 month | 1.65 | 57 | 64 | 4.03 | . |
| 179 | 10.58 | White/Caucasian | Y | Prednisone/Prednisolone | >=12 month | 1.51 | 68 | 64 | 2.63 | . |
| 182 | 10.53 | White/Caucasian | Y | Prednisone/Prednisolone | >=12 month | 2.7 | 88 | 89 | 5.23 | . |
| 184 | 10.34 | White/Caucasian | Y | Prednisone/Prednisolone | >=12 month | 1.6 | 87 | 80 | 2.56 | . |
| 187 | 10.25 | White/Caucasian | N | None | >=12 month | 1.6 | 63 | 73 | 3.05 | . |
| 208 | 10.03 | Asian | N | None | <12 month | 1.87 | 69 | 76 | 2.82 | . |

Listing 16.2.9.2 (Page 5 of 5)
Demographics Characteristics and Baseline Characteristics
(Matched Population for Age at FVC < 1 Liter Analysis)

Study: CINRG

| Subject ID | Baseline Age (yrs) | Race | Steroid Use | Baseline corticosteroid type | Cumul. BL Steroid Duration | Baseline FVC (L) | Baseline %-predicted FVC | Baseline %-predicted FEV1 | Baseline PEF | Baseline EK Score |
|------------|--------------------|-----------------|-------------|------------------------------|----------------------------|------------------|--------------------------|---------------------------|--------------|-------------------|
| 233 | 9.83 | White/Caucasian | N | None | <12 month | 1.78 | 70 | 79 | 3.41 | . |
| 245 | 9.05 | Asian | N | None | <12 month | 1.48 | 73 | 81 | 2.89 | . |

Listing 16.2.9.3 (Page 1 of 4)
 Demographics Characteristics and Baseline Characteristics
 (Matched Population for Age at Loss of Ambulation)

Study: 019

| Subject ID | Baseline Age (yrs) | Race | Steroid Use | Baseline corticosteroid type | Baseline time 10m run/walk (sec.) | Baseline time stand fr. supine (sec.) | Baseline NSAA total score |
|------------|--------------------|-----------------|-------------|------------------------------|-----------------------------------|---------------------------------------|---------------------------|
| 001013 | 15.21 | White/Caucasian | Y | Deflazacort | 8.7 | 30 | 16 |
| 003003 | 15.37 | Asian | Y | Deflazacort | 11.6 | . | 7 |
| 011001 | 14.91 | White/Caucasian | Y | Prednisone/Prednisolone | 11.1 | . | 6 |
| 011003 | 15.91 | White/Caucasian | Y | Prednisone/Prednisolone | 9.9 | . | 6 |
| 012005 | 13.63 | White/Caucasian | Y | Prednisone/Prednisolone | 12.1 | 30 | 7 |
| 021002 | 15.93 | White/Caucasian | Y | Deflazacort | 5.8 | . | 23 |
| 021003 | 13.59 | White/Caucasian | Y | Deflazacort | 4.8 | 3.6 | 29 |
| 021004 | 12.67 | White/Caucasian | Y | Deflazacort | 5.9 | 11.78 | 20 |
| 021005 | 12.62 | White/Caucasian | Y | Deflazacort | 4.2 | 5.5 | 26 |
| 031002 | 11.32 | White/Caucasian | Y | Deflazacort | 5.6 | 7.2 | 25 |
| 031004 | 13.46 | White/Caucasian | Y | Deflazacort | 4.9 | 4 | 19 |
| 033001 | 10.11 | White/Caucasian | Y | Deflazacort | 5.78 | 5.44 | 25 |
| 034001 | 13.48 | White/Caucasian | Y | Deflazacort | 21.4 | . | 3 |
| 034002 | 14 | Asian | Y | Deflazacort | 10.2 | . | 7 |
| 041001 | 15.06 | White/Caucasian | Y | Deflazacort | 7.4 | 6.4 | 23 |
| 041003 | 12.99 | White/Caucasian | Y | Prednisone/Prednisolone | 9.1 | . | 12 |
| 051004 | 12.12 | White/Caucasian | Y | Deflazacort | 10.3 | 5.2 | 29 |
| 061002 | 12.02 | White/Caucasian | Y | Deflazacort | 20.9 | . | 5 |
| 062002 | 12.04 | White/Caucasian | Y | Deflazacort | 8.46 | 30 | 16 |
| 062004 | 12.77 | White/Caucasian | Y | Deflazacort | 5.2 | 11.8 | 23 |
| 062006 | 13.77 | White/Caucasian | Y | Deflazacort | 5.4 | 7.2 | 28 |
| 062007 | 11.32 | Unknown | Y | Deflazacort | 3.54 | 3.44 | 32 |
| 063001 | 11 | White/Caucasian | Y | Deflazacort | 5.43 | 4.5 | 24 |
| 063003 | 9.96 | White/Caucasian | Y | Deflazacort | 4.32 | 4.2 | 32 |
| 063004 | 10.25 | White/Caucasian | Y | Prednisone/Prednisolone | 5.1 | 5.65 | 27 |

Listing 16.2.9.3 (Page 2 of 4)
 Demographics Characteristics and Baseline Characteristics
 (Matched Population for Age at Loss of Ambulation)

Study: 019

| Subject ID | Baseline Age (yrs) | Race | Steroid Use | Baseline corticosteroid type | Baseline time 10m run/walk (sec.) | Baseline time stand fr. supine (sec.) | Baseline NSAA total score |
|------------|--------------------|-----------------|-------------|------------------------------|-----------------------------------|---------------------------------------|---------------------------|
| 063005 | 9.43 | White/Caucasian | Y | Deflazacort | 5.16 | 4.7 | 29 |
| 071002 | 10.41 | White/Caucasian | Y | Prednisone/Prednisolone | 5.9 | 12 | 19 |
| 081001 | 12.89 | White/Caucasian | Y | Deflazacort | 9.2 | 10.4 | 13 |
| 082001 | 11.43 | White/Caucasian | Y | Prednisone/Prednisolone | 9.04 | 10.8 | 21 |
| 082002 | 11.16 | White/Caucasian | Y | Deflazacort | 5.5 | . | 26 |
| 091001 | 9.29 | White/Caucasian | Y | Prednisone/Prednisolone | 4.13 | 5.03 | 25 |
| 091003 | 9.14 | White/Caucasian | Y | Prednisone/Prednisolone | 11.27 | 30 | 11 |
| 091005 | 12.29 | White/Caucasian | Y | Prednisone/Prednisolone | 5.44 | 5.38 | 23 |
| 092001 | 13.17 | White/Caucasian | Y | Deflazacort | 17 | . | 8 |
| 101001 | 11.96 | White/Caucasian | N | None | 5.7 | 3.8 | 24 |
| 101002 | 12.75 | White/Caucasian | Y | Deflazacort | 7.4 | 8.3 | 21 |
| 101003 | 12.06 | White/Caucasian | Y | Prednisone/Prednisolone | 4.2 | 3.6 | 34 |
| 101005 | 14.52 | White/Caucasian | Y | Deflazacort | 8.4 | . | 10 |
| 101006 | 17.02 | White/Caucasian | Y | Prednisone/Prednisolone | 26.4 | 30 | 8 |
| 101009 | 13.67 | White/Caucasian | Y | Deflazacort | 13.4 | . | 6 |
| 101011 | 9.23 | White/Caucasian | Y | Prednisone/Prednisolone | 4 | 2.3 | 31 |
| 102001 | 12.82 | Asian | N | None | 7.32 | 14.56 | 25 |
| 103004 | 13.98 | White/Caucasian | Y | Prednisone/Prednisolone | 9.2 | 14.2 | 15 |
| 103006 | 13.48 | White/Caucasian | Y | Deflazacort | 7.1 | . | 23 |

Listing 16.2.9.3 (Page 3 of 4)
 Demographics Characteristics and Baseline Characteristics
 (Matched Population for Age at Loss of Ambulation)

Study: CINRG

| Subject ID | Baseline Age (yrs) | Race | Steroid Use | Baseline corticosteroid type | Baseline time 10m run/walk (sec.) | Baseline time stand fr. supine (sec.) | Baseline NSAA total score |
|------------|--------------------|-----------------|-------------|------------------------------|-----------------------------------|---------------------------------------|---------------------------|
| 075 | 16.7 | White/Caucasian | Y | Prednisone/Prednisolone | 8.31 | . | . |
| 080 | 16.75 | White/Caucasian | Y | Deflazacort | 12 | . | . |
| 085 | 15.71 | White/Caucasian | Y | Deflazacort | 7.16 | . | . |
| 089 | 15.67 | White/Caucasian | N | None | 12.13 | . | . |
| 094 | 15.08 | White/Caucasian | Y | Deflazacort | 10.84 | . | . |
| 104 | 13.55 | White/Caucasian | Y | Deflazacort | 8.8 | . | . |
| 107 | 13.44 | White/Caucasian | Y | Deflazacort | 9.43 | . | . |
| 110 | 13.96 | White/Caucasian | Y | Prednisone/Prednisolone | 10.5 | . | . |
| 114 | 14.14 | White/Caucasian | Y | Prednisone/Prednisolone | 6.5 | 9.01 | . |
| 120 | 13.18 | White/Caucasian | Y | Deflazacort | 11.62 | . | . |
| 122 | 13.48 | Asian | Y | Deflazacort | 13.37 | . | . |
| 127 | 13.82 | White/Caucasian | Y | Deflazacort | 4.04 | 2.71 | . |
| 128 | 13.18 | White/Caucasian | Y | Prednisone/Prednisolone | 7.09 | . | . |
| 130 | 13.17 | White/Caucasian | Y | Prednisone/Prednisolone | 6.38 | . | . |
| 135 | 12.31 | White/Caucasian | Y | Prednisone/Prednisolone | 11.19 | 27.68 | . |
| 141 | 12.09 | White/Caucasian | Y | Prednisone/Prednisolone | 8.15 | 7.25 | . |
| 143 | 13.82 | White/Caucasian | Y | Deflazacort | 6.45 | . | . |
| 144 | 12.23 | White/Caucasian | Y | Deflazacort | 13.47 | . | . |
| 145 | 12.43 | White/Caucasian | Y | Prednisone/Prednisolone | 4.28 | 6.34 | . |
| 146 | 12.34 | White/Caucasian | Y | Prednisone/Prednisolone | 11.5 | 12.28 | . |
| 147 | 12.26 | White/Caucasian | Y | Prednisone/Prednisolone | 7.71 | 12.73 | . |
| 148 | 11.83 | White/Caucasian | Y | Deflazacort | 8.13 | 14.9 | . |
| 149 | 11.21 | White/Caucasian | Y | Prednisone/Prednisolone | 6.64 | 5.62 | . |
| 151 | 11.68 | White/Caucasian | Y | Deflazacort | 7.84 | 11.9 | . |
| 154 | 12.07 | White/Caucasian | Y | Deflazacort | 11.41 | . | . |

Listing 16.2.9.3 (Page 4 of 4)
Demographics Characteristics and Baseline Characteristics
(Matched Population for Age at Loss of Ambulation)

Study: CINRG

| Subject ID | Baseline Age (yrs) | Race | Steroid Use | Baseline corticosteroid type | Baseline time 10m run/walk (sec.) | Baseline time stand fr. supine (sec.) | Baseline NSAA total score |
|------------|--------------------|------------------------|-------------|------------------------------|-----------------------------------|---------------------------------------|---------------------------|
| 156 | 11.78 | White/Caucasian | Y | Deflazacort | 6.81 | 11.3 | . |
| 161 | 11.43 | White/Caucasian | Y | Prednisone/Prednisolone | 5 | 5.29 | . |
| 165 | 11.06 | White/Caucasian | Y | Deflazacort | 12.38 | . | . |
| 167 | 10.42 | Black/African American | Y | Prednisone/Prednisolone | 12.52 | . | . |
| 178 | 10.11 | Asian | Y | Prednisone/Prednisolone | 10.4 | 10.23 | . |
| 181 | 10.52 | White/Caucasian | Y | Deflazacort | 3.62 | 3.78 | . |
| 186 | 10.33 | White/Caucasian | Y | Deflazacort | 12.1 | . | . |
| 188 | 11.07 | Pacific Islander | Y | Deflazacort | 4.63 | 4.43 | . |
| 190 | 9.73 | White/Caucasian | Y | Prednisone/Prednisolone | 11.91 | . | . |
| 191 | 10.18 | Other | Y | Deflazacort | 9.81 | 18.66 | . |
| 193 | 10.38 | White/Caucasian | Y | Deflazacort | 5.53 | 7.82 | . |
| 194 | 9.8 | White/Caucasian | Y | Prednisone/Prednisolone | 8.19 | 14.42 | . |
| 195 | 11.47 | Other | Y | Prednisone/Prednisolone | 12 | 13.91 | . |
| 196 | 9.97 | White/Caucasian | Y | Deflazacort | 4.81 | 5.59 | . |
| 197 | 9.13 | Asian | Y | Prednisone/Prednisolone | 10.06 | 7.03 | . |
| 199 | 9.28 | White/Caucasian | Y | Deflazacort | 5.84 | 6.84 | . |
| 205 | 9.38 | White/Caucasian | Y | Deflazacort | 22.63 | . | . |
| 206 | 9.86 | White/Caucasian | Y | Deflazacort | 6.15 | 20.15 | . |
| 211 | 9.28 | Asian | Y | Deflazacort | 8.88 | 12 | . |
| 228 | 9.34 | White/Caucasian | Y | Prednisone/Prednisolone | 9.5 | . | . |
| 249 | 9.19 | Asian | Y | Prednisone/Prednisolone | 9.27 | 7.86 | . |

Listing 16.2.9.4 (Page 1 of 32)
Listing of FVC
(Matched Population for Piece-wise Regression Analysis on FVC)

Study: 019

| Subject ID | Visit Year | Age(yrs) at Visit | Ambulatory Status at Visit | Steroid Use | Steroid Use 24 Month | FVC (L) |
|------------|------------|-------------------|----------------------------|-------------|----------------------|---------|
| 001013 | 2015 | 17.05 | 0 | 1 | >=24 mo | 2.56 |
| | 2016 | 17.51 | 0 | 1 | >=24 mo | 2.25 |
| | 2016 | 17.97 | 0 | 1 | >=24 mo | 2.14 |
| | 2017 | 18.6 | 0 | 1 | >=24 mo | 1.51 |
| 003003 | 2013 | 16.27 | 0 | 1 | >=24 mo | 2.26 |
| | 2014 | 16.73 | 0 | 1 | >=24 mo | 2.02 |
| | 2014 | 17.21 | 0 | 1 | >=24 mo | 2.11 |
| | 2015 | 17.65 | 0 | 1 | >=24 mo | 2.28 |
| | 2015 | 18.13 | 0 | 1 | >=24 mo | 2.08 |
| | 2016 | 18.59 | 0 | 1 | >=24 mo | 2.21 |
| | 2016 | 19.07 | 0 | 1 | >=24 mo | 2.3 |
| | 2016 | 19.51 | 0 | 1 | >=24 mo | 2.67 |
| 011001 | 2015 | 16.29 | 0 | 1 | >=24 mo | 1.52 |
| | 2016 | 17.68 | 0 | 1 | >=24 mo | 1.47 |
| | 2016 | 18.12 | 0 | 1 | >=24 mo | 1.41 |
| | 2017 | 18.59 | 0 | 1 | >=24 mo | 1.23 |
| 011002 | 2013 | 14.05 | 0 | 0 | >=24 mo | 2.57 |
| 011003 | 2015 | 17.3 | 0 | 1 | >=24 mo | 2.73 |

Note: Ambulatory status: 1=ambulatory, 0=non-ambulatory. Steroid Use: 1=Yes, 0=No.

Listing 16.2.9.4 (Page 2 of 32)
 Listing of FVC
 (Matched Population for Piece-wise Regression Analysis on FVC)

Study: 019

| Subject ID | Visit Year | Age(yrs) at Visit | Ambulatory Status at Visit | Steroid Use | Steroid Use 24 Month | FVC (L) |
|------------|------------|-------------------|----------------------------|-------------|----------------------|---------|
| 011003 | 2015 | 17.75 | 0 | 1 | >=24 mo | 1.96 |
| | 2016 | 18.66 | 0 | 1 | >=24 mo | 1.76 |
| | 2016 | 19.14 | 0 | 1 | >=24 mo | 1.61 |
| | 2017 | 19.59 | 0 | 1 | >=24 mo | 1.31 |
| 011004 | 2014 | 15.94 | 0 | 1 | >=24 mo | 2.4 |
| | 2015 | 16.87 | 0 | 1 | >=24 mo | 3.07 |
| | 2015 | 17.33 | 0 | 1 | >=24 mo | 1.52 |
| | 2015 | 17.78 | 0 | 1 | >=24 mo | 1.58 |
| | 2016 | 18.25 | 0 | 1 | >=24 mo | 1.32 |
| | 2016 | 18.7 | 0 | 1 | >=24 mo | 1.3 |
| | 2017 | 19.16 | 0 | 1 | >=24 mo | 1.28 |
| 011005 | 2013 | 13.4 | 0 | 1 | >=24 mo | 2.22 |
| | 2014 | 13.85 | 0 | 1 | >=24 mo | 2.3 |
| | 2015 | 14.78 | 0 | 1 | >=24 mo | 2.69 |
| | 2015 | 15.26 | 0 | 1 | >=24 mo | 1.31 |
| | 2016 | 16.18 | 0 | 1 | >=24 mo | 1.48 |
| | 2016 | 16.62 | 0 | 1 | >=24 mo | 1.34 |
| | 2017 | 17.09 | 0 | 1 | >=24 mo | 1.3 |

Note: Ambulatory status: 1=ambulatory, 0=non-ambulatory. Steroid Use: 1=Yes, 0=No.

Listing 16.2.9.4 (Page 3 of 32)
Listing of FVC
(Matched Population for Piece-wise Regression Analysis on FVC)

Study: 019

| Subject ID | Visit Year | Age(yrs) at Visit | Ambulatory Status at Visit | Steroid Use | Steroid Use 24 Month | FVC (L) |
|------------|------------|-------------------|----------------------------|-------------|----------------------|---------|
| 011006 | 2016 | 15.8 | 0 | 1 | >=24 mo | 1.21 |
| | 2016 | 16.25 | 0 | 1 | >=24 mo | 0.94 |
| | 2017 | 16.71 | 0 | 1 | >=24 mo | 1.65 |
| 012001 | 2013 | 15.73 | 0 | 1 | >=24 mo | 2.14 |
| | 2014 | 16.19 | 0 | 1 | >=24 mo | 2.17 |
| | 2014 | 16.65 | 0 | 1 | >=24 mo | 2.25 |
| | 2015 | 17.13 | 0 | 1 | >=24 mo | 2.22 |
| | 2015 | 17.58 | 0 | 1 | >=24 mo | 2.12 |
| | 2015 | 18.03 | 0 | 1 | >=24 mo | 2.12 |
| | 2016 | 18.51 | 0 | 1 | >=24 mo | 2.05 |
| | 2016 | 18.98 | 0 | 1 | >=24 mo | 1.88 |
| | 2017 | 19.43 | 0 | 1 | >=24 mo | 1.9 |
| 012005 | 2015 | 15.49 | 0 | 1 | >=24 mo | 1.48 |
| | 2016 | 15.97 | 0 | 1 | >=24 mo | 1.51 |
| | 2016 | 16.44 | 0 | 1 | >=24 mo | 1.6 |
| | 2017 | 17.02 | 0 | 1 | >=24 mo | 1.76 |
| 021001 | 2012 | 14.77 | 0 | 1 | >=24 mo | 2.09 |
| | 2012 | 15.2 | 0 | 1 | >=24 mo | 2.05 |

Note: Ambulatory status: 1=ambulatory, 0=non-ambulatory. Steroid Use: 1=Yes, 0=No.

Listing 16.2.9.4 (Page 4 of 32)
Listing of FVC
(Matched Population for Piece-wise Regression Analysis on FVC)

Study: 019

| Subject ID | Visit Year | Age(yrs) at Visit | Ambulatory Status at Visit | Steroid Use | Steroid Use 24 Month | FVC (L) |
|------------|------------|-------------------|----------------------------|-------------|----------------------|---------|
| 021001 | 2013 | 15.66 | 0 | 1 | >=24 mo | 2.03 |
| | 2013 | 16.12 | 0 | 1 | >=24 mo | 2.15 |
| | 2014 | 16.6 | 0 | 1 | >=24 mo | 1.76 |
| | 2014 | 17.04 | 0 | 1 | >=24 mo | 1.44 |
| | 2015 | 17.52 | 0 | 1 | >=24 mo | 1.32 |
| | 2015 | 17.98 | 0 | 1 | >=24 mo | 1.4 |
| | 2016 | 18.44 | 0 | 1 | >=24 mo | 1.42 |
| | 2016 | 18.9 | 0 | 1 | >=24 mo | 1.53 |
| | 2017 | 19.6 | 0 | 0 | >=24 mo | 1.52 |
| 021004 | 2016 | 16.35 | 0 | 1 | >=24 mo | 0.85 |
| | 2016 | 16.8 | 0 | 1 | >=24 mo | 0.94 |
| | 2017 | 17.48 | 0 | 0 | >=24 mo | 0.89 |
| 021005 | 2017 | 17.44 | 0 | 0 | >=24 mo | 2.24 |
| 031001 | 2012 | 21.02 | 0 | 1 | >=24 mo | 1.16 |
| | 2013 | 21.5 | 0 | 1 | >=24 mo | 1.1 |
| | 2013 | 21.94 | 0 | 1 | >=24 mo | 1.04 |
| | 2014 | 22.41 | 0 | 1 | >=24 mo | 0.6 |
| | 2015 | 23.34 | 0 | 1 | >=24 mo | 0.89 |

Note: Ambulatory status: 1=ambulatory, 0=non-ambulatory. Steroid Use: 1=Yes, 0=No.

Listing 16.2.9.4 (Page 5 of 32)
Listing of FVC
(Matched Population for Piece-wise Regression Analysis on FVC)

Study: 019

| Subject ID | Visit Year | Age(yrs) at Visit | Ambulatory Status at Visit | Steroid Use | Steroid Use 24 Month | FVC (L) |
|------------|------------|-------------------|----------------------------|-------------|----------------------|---------|
| 031001 | 2015 | 23.78 | 0 | 1 | >=24 mo | 0.78 |
| | 2016 | 24.24 | 0 | 1 | >=24 mo | 1.07 |
| | 2016 | 24.73 | 0 | 1 | >=24 mo | 0.93 |
| 031002 | 2016 | 15 | 0 | 0 | >=24 mo | 1.6 |
| 031003 | 2012 | 11.78 | 0 | 1 | >=24 mo | 1.31 |
| | 2013 | 12.67 | 0 | 1 | >=24 mo | 1.26 |
| | 2013 | 13.17 | 0 | 1 | >=24 mo | 1.1 |
| | 2014 | 13.67 | 0 | 1 | >=24 mo | 0.9 |
| | 2014 | 14.09 | 0 | 1 | >=24 mo | 0.93 |
| | 2015 | 15.01 | 0 | 1 | >=24 mo | 0.65 |
| | 2016 | 15.47 | 0 | 1 | >=24 mo | 0.74 |
| | 2016 | 15.93 | 0 | 1 | >=24 mo | 0.72 |
| 034001 | 2013 | 13.94 | 0 | 1 | >=24 mo | 1.29 |
| | 2013 | 14.38 | 0 | 1 | >=24 mo | 1.36 |
| | 2014 | 14.86 | 0 | 1 | >=24 mo | 1.36 |
| | 2014 | 15.32 | 0 | 1 | >=24 mo | 1.39 |
| | 2015 | 15.8 | 0 | 1 | >=24 mo | 1.53 |
| | 2015 | 16.24 | 0 | 1 | >=24 mo | 1.58 |

Note: Ambulatory status: 1=ambulatory, 0=non-ambulatory. Steroid Use: 1=Yes, 0=No.

Listing 16.2.9.4 (Page 6 of 32)
Listing of FVC
(Matched Population for Piece-wise Regression Analysis on FVC)

Study: 019

| Subject ID | Visit Year | Age(yrs) at Visit | Ambulatory Status at Visit | Steroid Use | Steroid Use 24 Month | FVC (L) |
|------------|------------|-------------------|----------------------------|-------------|----------------------|---------|
| 034001 | 2015 | 16.68 | 0 | 1 | >=24 mo | 1.39 |
| | 2016 | 17.16 | 0 | 1 | >=24 mo | 1.22 |
| | 2016 | 17.62 | 0 | 1 | >=24 mo | 1.04 |
| 034002 | 2013 | 14.9 | 0 | 1 | >=24 mo | 1.83 |
| | 2014 | 15.38 | 0 | 1 | >=24 mo | 1.91 |
| | 2014 | 15.84 | 0 | 1 | >=24 mo | 1.72 |
| | 2015 | 16.3 | 0 | 1 | >=24 mo | 2.12 |
| | 2015 | 16.76 | 0 | 1 | >=24 mo | 2.11 |
| | 2015 | 17.2 | 0 | 1 | >=24 mo | 1.97 |
| | 2016 | 17.66 | 0 | 1 | >=24 mo | 1.74 |
| | 2016 | 18.14 | 0 | 1 | >=24 mo | 1.79 |
| 041004 | 2012 | 13.59 | 0 | 1 | >=24 mo | 1.83 |
| | 2013 | 14.06 | 0 | 1 | >=24 mo | 2.01 |
| | 2013 | 14.52 | 0 | 1 | >=24 mo | 2.02 |
| | 2014 | 14.98 | 0 | 1 | >=24 mo | 1.88 |
| | 2014 | 15.42 | 0 | 1 | >=24 mo | 2.13 |
| | 2015 | 15.88 | 0 | 1 | >=24 mo | 2.06 |
| | 2015 | 16.36 | 0 | 1 | >=24 mo | 2.45 |

Note: Ambulatory status: 1=ambulatory, 0=non-ambulatory. Steroid Use: 1=Yes, 0=No.

Listing 16.2.9.4 (Page 7 of 32)
Listing of FVC
(Matched Population for Piece-wise Regression Analysis on FVC)

Study: 019

| Subject ID | Visit Year | Age(yrs) at Visit | Ambulatory Status at Visit | Steroid Use | Steroid Use 24 Month | FVC (L) |
|------------|------------|-------------------|----------------------------|-------------|----------------------|---------|
| 041004 | 2016 | 16.84 | 0 | 1 | >=24 mo | 2.44 |
| | 2016 | 17.25 | 0 | 1 | >=24 mo | 2.47 |
| | 2017 | 17.72 | 0 | 1 | >=24 mo | 2.5 |
| | 2017 | 18.19 | 0 | 1 | >=24 mo | 2.1 |
| 041005 | 2015 | 15.01 | 0 | 1 | >=24 mo | 1.66 |
| | 2015 | 15.46 | 0 | 1 | >=24 mo | 1.56 |
| | 2016 | 15.93 | 0 | 1 | >=24 mo | 1.54 |
| | 2016 | 16.35 | 0 | 1 | >=24 mo | 1.41 |
| | 2017 | 16.85 | 0 | 1 | >=24 mo | 1.32 |
| | 2017 | 17.29 | 0 | 1 | >=24 mo | 1.27 |
| 042001 | 2012 | 11.03 | 0 | 1 | >=24 mo | 1.71 |
| | 2013 | 11.47 | 0 | 1 | >=24 mo | 2.33 |
| | 2013 | 11.93 | 0 | 1 | >=24 mo | 2.02 |
| | 2014 | 12.92 | 0 | 1 | >=24 mo | 2.35 |
| | 2016 | 14.69 | 0 | 1 | >=24 mo | 3.86 |
| | 2017 | 15.18 | 0 | 1 | >=24 mo | 2.79 |
| | 2017 | 15.64 | 0 | 0 | >=24 mo | 2.27 |
| 042002 | 2012 | 12.65 | 0 | 1 | >=24 mo | 2.42 |

Note: Ambulatory status: 1=ambulatory, 0=non-ambulatory. Steroid Use: 1=Yes, 0=No.

Listing 16.2.9.4 (Page 8 of 32)
 Listing of FVC
 (Matched Population for Piece-wise Regression Analysis on FVC)

Study: 019

| Subject ID | Visit Year | Age(yrs) at Visit | Ambulatory Status at Visit | Steroid Use | Steroid Use 24 Month | FVC (L) |
|------------|------------|-------------------|----------------------------|-------------|----------------------|---------|
| 042002 | 2013 | 13.1 | 0 | 1 | >=24 mo | 2.37 |
| | 2013 | 13.56 | 0 | 1 | >=24 mo | 2.59 |
| | 2014 | 14.51 | 0 | 1 | >=24 mo | 2.61 |
| | 2015 | 15.45 | 0 | 1 | >=24 mo | 2.57 |
| | 2016 | 16.31 | 0 | 1 | >=24 mo | 2.34 |
| | 2017 | 16.81 | 0 | 1 | >=24 mo | 2.7 |
| | 2017 | 17.27 | 0 | 1 | >=24 mo | 2.25 |
| 043001 | 2012 | 12.29 | 0 | 1 | >=24 mo | 2.01 |
| | 2013 | 12.74 | 0 | 1 | >=24 mo | 2.16 |
| | 2013 | 13.19 | 0 | 0 | >=24 mo | 2.22 |
| | 2014 | 13.65 | 0 | 0 | >=24 mo | 2.3 |
| | 2014 | 14.14 | 0 | 0 | >=24 mo | 2.51 |
| | 2015 | 14.61 | 0 | 0 | >=24 mo | 2.51 |
| 043002 | 2012 | 14.36 | 0 | 1 | >=24 mo | 2.54 |
| | 2013 | 14.81 | 0 | 1 | >=24 mo | 2.41 |
| | 2013 | 15.28 | 0 | 1 | >=24 mo | 2.2 |
| | 2014 | 15.75 | 0 | 1 | >=24 mo | 2.32 |
| | 2014 | 16.22 | 0 | 1 | >=24 mo | 2.25 |

Note: Ambulatory status: 1=ambulatory, 0=non-ambulatory. Steroid Use: 1=Yes, 0=No.

Listing 16.2.9.4 (Page 9 of 32)
 Listing of FVC
 (Matched Population for Piece-wise Regression Analysis on FVC)

Study: 019

| Subject ID | Visit Year | Age(yrs) at Visit | Ambulatory Status at Visit | Steroid Use | Steroid Use 24 Month | FVC (L) |
|------------|------------|-------------------|----------------------------|-------------|----------------------|---------|
| 043002 | 2015 | 16.67 | 0 | 1 | >=24 mo | 2.26 |
| | 2015 | 17.11 | 0 | 1 | >=24 mo | 2.07 |
| | 2016 | 17.6 | 0 | 1 | >=24 mo | 2.22 |
| | 2016 | 18.08 | 0 | 1 | >=24 mo | 2.06 |
| | 2016 | 18.51 | 0 | 1 | >=24 mo | 1.83 |
| | 2016 | 18.51 | 0 | 1 | >=24 mo | 1.83 |
| | 2017 | 18.97 | 0 | 1 | >=24 mo | 1.96 |
| 051001 | 2013 | 20.02 | 0 | 1 | >=24 mo | 1.16 |
| | 2014 | 20.53 | 0 | 1 | >=24 mo | 1 |
| | 2014 | 20.93 | 0 | 1 | >=24 mo | 0.78 |
| | 2015 | 21.39 | 0 | 1 | >=24 mo | 0.81 |
| | 2015 | 21.84 | 0 | 1 | >=24 mo | 0.87 |
| | 2016 | 22.76 | 0 | 1 | >=24 mo | 0.8 |
| | 2016 | 23.21 | 0 | 1 | >=24 mo | 0.73 |
| | 2017 | 23.69 | 0 | 1 | >=24 mo | 0.74 |
| | 2017 | 24.15 | 0 | 1 | >=24 mo | 0.77 |
| 051003 | 2013 | 15.87 | 0 | 1 | >=24 mo | 2.44 |
| | 2014 | 16.32 | 0 | 1 | >=24 mo | 2.41 |
| | 2014 | 16.78 | 0 | 1 | >=24 mo | 2.23 |

Note: Ambulatory status: 1=ambulatory, 0=non-ambulatory. Steroid Use: 1=Yes, 0=No.

Listing 16.2.9.4 (Page 10 of 32)
Listing of FVC
(Matched Population for Piece-wise Regression Analysis on FVC)

Study: 019

| Subject ID | Visit Year | Age(yrs) at Visit | Ambulatory Status at Visit | Steroid Use | Steroid Use 24 Month | FVC (L) |
|------------|------------|-------------------|----------------------------|-------------|----------------------|---------|
| 051003 | 2015 | 17.24 | 0 | 1 | >=24 mo | 2.22 |
| | 2015 | 17.69 | 0 | 1 | >=24 mo | 2.1 |
| | 2016 | 18.61 | 0 | 1 | >=24 mo | 1.79 |
| | 2016 | 19.06 | 0 | 1 | >=24 mo | 1.56 |
| | 2017 | 19.53 | 0 | 1 | >=24 mo | 1.37 |
| | 2017 | 20.01 | 0 | 1 | >=24 mo | 1.34 |
| | 052006 | 2012 | 11.48 | 0 | 1 | >=24 mo |
| 2013 | | 11.93 | 0 | 1 | >=24 mo | 2.11 |
| 2013 | | 12.4 | 0 | 1 | >=24 mo | 2.04 |
| 2014 | | 12.87 | 0 | 1 | >=24 mo | 2.48 |
| 2014 | | 13.3 | 0 | 1 | >=24 mo | 2.26 |
| 2015 | | 13.8 | 0 | 1 | >=24 mo | 1.9 |
| 2015 | | 14.26 | 0 | 1 | >=24 mo | 2.05 |
| 061001 | 2013 | 12 | 0 | 1 | >=24 mo | 1.93 |
| | 2013 | 12.46 | 0 | 1 | >=24 mo | 2 |
| | 2014 | 12.93 | 0 | 1 | >=24 mo | 1.88 |
| | 2014 | 13.38 | 0 | 0 | >=24 mo | 2.15 |
| | 2015 | 13.84 | 0 | 0 | >=24 mo | 2.18 |

Note: Ambulatory status: 1=ambulatory, 0=non-ambulatory. Steroid Use: 1=Yes, 0=No.

Listing 16.2.9.4 (Page 11 of 32)
Listing of FVC
(Matched Population for Piece-wise Regression Analysis on FVC)

Study: 019

| Subject ID | Visit Year | Age(yrs) at Visit | Ambulatory Status at Visit | Steroid Use | Steroid Use 24 Month | FVC (L) |
|------------|------------|-------------------|----------------------------|-------------|----------------------|---------|
| 061001 | 2015 | 14.31 | 0 | 0 | >=24 mo | 2.48 |
| | 2016 | 14.78 | 0 | 0 | >=24 mo | 2.2 |
| | 2016 | 15.21 | 0 | 0 | >=24 mo | 2.76 |
| | 2017 | 15.69 | 0 | 0 | >=24 mo | 2.88 |
| | 2017 | 16.13 | 0 | 0 | >=24 mo | 2.85 |
| | 2017 | 16.61 | 0 | 0 | >=24 mo | 2.6 |
| 061002 | 2013 | 12.48 | 0 | 1 | >=24 mo | 1.94 |
| | 2014 | 12.95 | 0 | 1 | >=24 mo | 1.67 |
| | 2014 | 13.4 | 0 | 1 | >=24 mo | 1.95 |
| | 2015 | 13.86 | 0 | 1 | >=24 mo | 1.3 |
| | 2015 | 14.34 | 0 | 1 | >=24 mo | 2.02 |
| | 2016 | 14.78 | 0 | 1 | >=24 mo | 1.46 |
| | 2016 | 15.24 | 0 | 1 | >=24 mo | 1.93 |
| | 2017 | 15.72 | 0 | 1 | >=24 mo | 1.95 |
| | 2017 | 16.17 | 0 | 1 | >=24 mo | 2.16 |
| | 2017 | 16.64 | 0 | 0 | >=24 mo | 2.17 |
| 062001 | 2013 | 14.27 | 0 | 1 | >=24 mo | 2.52 |
| | 2013 | 14.74 | 0 | 1 | >=24 mo | 2.61 |

Note: Ambulatory status: 1=ambulatory, 0=non-ambulatory. Steroid Use: 1=Yes, 0=No.

Listing 16.2.9.4 (Page 12 of 32)
 Listing of FVC
 (Matched Population for Piece-wise Regression Analysis on FVC)

Study: 019

| Subject ID | Visit Year | Age (yrs) at Visit | Ambulatory Status at Visit | Steroid Use | Steroid Use 24 Month | FVC (L) |
|------------|------------|--------------------|----------------------------|-------------|----------------------|---------|
| 062001 | 2014 | 15.2 | 0 | 1 | >=24 mo | 2.75 |
| | 2014 | 15.64 | 0 | 1 | >=24 mo | 2.9 |
| | 2014 | 16.11 | 0 | 0 | >=24 mo | 2.62 |
| | 2015 | 16.57 | 0 | 0 | >=24 mo | 2.52 |
| | 2015 | 17.03 | 0 | 0 | >=24 mo | 2.29 |
| | 2016 | 17.49 | 0 | 1 | >=24 mo | 2.24 |
| | 2016 | 17.95 | 0 | 1 | >=24 mo | 2.01 |
| | 2017 | 18.41 | 0 | 1 | >=24 mo | 1.9 |
| 062002 | 2016 | 15.7 | 0 | 1 | >=24 mo | 1.94 |
| | 2017 | 16.16 | 0 | 1 | >=24 mo | 1.94 |
| | 2017 | 16.62 | 0 | 0 | >=24 mo | 2.39 |
| 062004 | 2015 | 15.53 | 0 | 1 | >=24 mo | 2.08 |
| | 2016 | 15.99 | 0 | 1 | >=24 mo | 2.04 |
| | 2016 | 16.44 | 0 | 1 | >=24 mo | 2.12 |
| 063001 | 2017 | 15.16 | 0 | 1 | >=24 mo | 1.97 |
| | 2017 | 15.6 | 0 | 0 | >=24 mo | 1.96 |
| 063002 | 2013 | 13.21 | 0 | 1 | >=24 mo | 2.48 |
| | 2013 | 13.65 | 0 | 1 | >=24 mo | 2.48 |

Note: Ambulatory status: 1=ambulatory, 0=non-ambulatory. Steroid Use: 1=Yes, 0=No.

Listing 16.2.9.4 (Page 13 of 32)
Listing of FVC
(Matched Population for Piece-wise Regression Analysis on FVC)

Study: 019

| Subject ID | Visit Year | Age(yrs) at Visit | Ambulatory Status at Visit | Steroid Use | Steroid Use 24 Month | FVC (L) |
|------------|------------|-------------------|----------------------------|-------------|----------------------|---------|
| 063005 | 2016 | 12.64 | 0 | 1 | >=24 mo | 1.89 |
| | 2016 | 13.12 | 0 | 1 | >=24 mo | 1.8 |
| | 2017 | 13.62 | 0 | 1 | >=24 mo | 1.65 |
| | 2017 | 14.02 | 0 | 0 | >=24 mo | 1.61 |
| 063006 | 2013 | 9.41 | 0 | 1 | >=24 mo | 1.53 |
| | 2013 | 9.87 | 0 | 1 | >=24 mo | 1.79 |
| | 2014 | 10.36 | 0 | 1 | >=24 mo | 1.85 |
| 071001 | 2013 | 12.63 | 0 | 1 | >=24 mo | 1.67 |
| | 2013 | 13.11 | 0 | 1 | >=24 mo | 1.96 |
| | 2013 | 13.52 | 0 | 1 | >=24 mo | 2 |
| | 2014 | 14 | 0 | 1 | >=24 mo | 1.94 |
| | 2014 | 14.48 | 0 | 1 | >=24 mo | 2 |
| | 2015 | 15.07 | 0 | 1 | >=24 mo | 1.9 |
| | 2015 | 15.42 | 0 | 1 | >=24 mo | 1.67 |
| | 2016 | 15.89 | 0 | 1 | >=24 mo | 1.76 |
| | 2016 | 16.35 | 0 | 1 | >=24 mo | 1.69 |
| | 2017 | 16.79 | 0 | 1 | >=24 mo | 1.55 |
| 2017 | 17.25 | 0 | 1 | >=24 mo | 1.5 | |

Note: Ambulatory status: 1=ambulatory, 0=non-ambulatory. Steroid Use: 1=Yes, 0=No.

Listing 16.2.9.4 (Page 14 of 32)
 Listing of FVC
 (Matched Population for Piece-wise Regression Analysis on FVC)

Study: 019

| Subject ID | Visit Year | Age(yrs) at Visit | Ambulatory Status at Visit | Steroid Use | Steroid Use 24 Month | FVC (L) |
|------------|------------|-------------------|----------------------------|-------------|----------------------|---------|
| 071002 | 2015 | 13.14 | 0 | 1 | >=24 mo | 1.81 |
| | 2016 | 13.64 | 0 | 1 | >=24 mo | 1.67 |
| | 2016 | 14.09 | 0 | 1 | >=24 mo | 1.8 |
| | 2017 | 14.53 | 0 | 1 | >=24 mo | 1.85 |
| | 2017 | 14.99 | 0 | 1 | >=24 mo | 1.93 |
| 081001 | 2014 | 14.26 | 0 | 1 | >=24 mo | 1.66 |
| | 2014 | 14.74 | 0 | 1 | >=24 mo | 1.65 |
| | 2015 | 15.17 | 0 | 1 | >=24 mo | 1.47 |
| | 2015 | 15.64 | 0 | 1 | >=24 mo | 1.37 |
| | 2016 | 16.1 | 0 | 1 | >=24 mo | 1.28 |
| | 2016 | 16.57 | 0 | 1 | >=24 mo | 1.13 |
| | 2017 | 17.02 | 0 | 1 | >=24 mo | 1.45 |
| 081002 | 2012 | 10.22 | 0 | 1 | >=24 mo | 1.79 |
| | 2013 | 10.67 | 0 | 1 | >=24 mo | 2.02 |
| | 2013 | 11.14 | 0 | 1 | >=24 mo | 1.98 |
| | 2014 | 11.6 | 0 | 1 | >=24 mo | 1.96 |
| | 2014 | 12.08 | 0 | 1 | >=24 mo | 2.03 |

Note: Ambulatory status: 1=ambulatory, 0=non-ambulatory. Steroid Use: 1=Yes, 0=No.

Listing 16.2.9.4 (Page 15 of 32)
Listing of FVC
(Matched Population for Piece-wise Regression Analysis on FVC)

Study: 019

| Subject ID | Visit Year | Age(yrs) at Visit | Ambulatory Status at Visit | Steroid Use | Steroid Use 24 Month | FVC (L) |
|------------|------------|-------------------|----------------------------|-------------|----------------------|---------|
| 081002 | 2015 | 12.5 | 0 | 1 | >=24 mo | 2.03 |
| | 2015 | 12.97 | 0 | 1 | >=24 mo | 2.01 |
| | 2016 | 13.43 | 0 | 1 | >=24 mo | 1.96 |
| | 2016 | 13.91 | 0 | 1 | >=24 mo | 2.18 |
| | 2017 | 14.45 | 0 | 0 | >=24 mo | 2.3 |
| 082003 | 2013 | 14.05 | 0 | 1 | >=24 mo | 2.38 |
| | 2013 | 14.51 | 0 | 1 | >=24 mo | 2.51 |
| | 2013 | 14.99 | 0 | 1 | >=24 mo | 2.38 |
| | 2014 | 15.43 | 0 | 1 | >=24 mo | 2.71 |
| | 2015 | 16.33 | 0 | 1 | >=24 mo | 3.42 |
| | 2015 | 16.83 | 0 | 1 | >=24 mo | 2.67 |
| | 2016 | 17.28 | 0 | 1 | >=24 mo | 2.61 |
| | 2016 | 17.74 | 0 | 1 | >=24 mo | 2.53 |
| 2017 | 18.28 | 0 | 0 | >=24 mo | 2.1 | |
| 082004 | 2013 | 14.33 | 0 | 1 | >=24 mo | 2.45 |
| | 2013 | 14.87 | 0 | 1 | >=24 mo | 2.55 |
| 091001 | 2016 | 12.99 | 0 | 1 | >=24 mo | 1.83 |
| | 2016 | 13.48 | 0 | 1 | >=24 mo | 1.78 |

Note: Ambulatory status: 1=ambulatory, 0=non-ambulatory. Steroid Use: 1=Yes, 0=No.

Listing 16.2.9.4 (Page 16 of 32)
Listing of FVC
(Matched Population for Piece-wise Regression Analysis on FVC)

Study: 019

| Subject ID | Visit Year | Age(yrs) at Visit | Ambulatory Status at Visit | Steroid Use | Steroid Use 24 Month | FVC (L) |
|------------|------------|-------------------|----------------------------|-------------|----------------------|---------|
| 091001 | 2017 | 13.74 | 0 | 1 | >=24 mo | 1.8 |
| 091002 | 2012 | 18.4 | 0 | 1 | >=24 mo | 2.58 |
| | 2013 | 18.84 | 0 | 1 | >=24 mo | 2.58 |
| | 2014 | 19.74 | 0 | 1 | >=24 mo | 2.85 |
| | 2014 | 20.17 | 0 | 1 | >=24 mo | 2.62 |
| | 2014 | 20.63 | 0 | 1 | >=24 mo | 2.46 |
| | 2015 | 21.16 | 0 | 1 | >=24 mo | 2.68 |
| | 2015 | 21.6 | 0 | 1 | >=24 mo | 2.7 |
| | 2016 | 22.07 | 0 | 1 | >=24 mo | 2.69 |
| | 2016 | 22.54 | 0 | 1 | >=24 mo | 2.6 |
| | 2017 | 22.77 | 0 | 1 | >=24 mo | 2.46 |
| 091003 | 2015 | 11.89 | 0 | 1 | >=24 mo | 1.12 |
| | 2015 | 12.37 | 0 | 1 | >=24 mo | 1.13 |
| | 2016 | 12.81 | 0 | 1 | >=24 mo | 1.07 |
| | 2016 | 13.31 | 0 | 1 | >=24 mo | 0.66 |
| | 2017 | 13.54 | 0 | 1 | >=24 mo | 0.51 |
| 091004 | 2012 | 15.27 | 0 | 1 | >=24 mo | 1.09 |
| | 2013 | 15.72 | 0 | 1 | >=24 mo | 1.17 |

Note: Ambulatory status: 1=ambulatory, 0=non-ambulatory. Steroid Use: 1=Yes, 0=No.

Listing 16.2.9.4 (Page 17 of 32)
 Listing of FVC
 (Matched Population for Piece-wise Regression Analysis on FVC)

Study: 019

| Subject ID | Visit Year | Age(yrs) at Visit | Ambulatory Status at Visit | Steroid Use | Steroid Use 24 Month | FVC (L) |
|------------|------------|-------------------|----------------------------|-------------|----------------------|---------|
| 091004 | 2014 | 16.62 | 0 | 1 | >=24 mo | 1.1 |
| | 2014 | 17.07 | 0 | 1 | >=24 mo | 1.12 |
| | 2014 | 17.52 | 0 | 1 | >=24 mo | 1.1 |
| | 2015 | 18.02 | 0 | 1 | >=24 mo | 0.98 |
| | 2015 | 18.49 | 0 | 1 | >=24 mo | 1 |
| | 2016 | 18.96 | 0 | 1 | >=24 mo | 0.79 |
| | 2016 | 19.43 | 0 | 1 | >=24 mo | 0.94 |
| | 2017 | 19.66 | 0 | 1 | >=24 mo | 0.94 |
| 092001 | 2015 | 15.03 | 0 | 1 | >=24 mo | 1.12 |
| | 2015 | 15.47 | 0 | 1 | >=24 mo | 2.07 |
| | 2016 | 16.85 | 0 | 1 | >=24 mo | 1 |
| 092002 | 2013 | 15.73 | 0 | 1 | >=24 mo | 1.45 |
| | 2013 | 16.18 | 0 | 1 | >=24 mo | 1.44 |
| | 2014 | 17.1 | 0 | 1 | >=24 mo | 1.38 |
| | 2015 | 17.59 | 0 | 1 | >=24 mo | 1.75 |
| | 2015 | 18.04 | 0 | 1 | >=24 mo | 1.98 |
| 092003 | 2013 | 14.28 | 0 | 1 | >=24 mo | 1.46 |
| | 2013 | 14.72 | 0 | 1 | >=24 mo | 1.6 |

Note: Ambulatory status: 1=ambulatory, 0=non-ambulatory. Steroid Use: 1=Yes, 0=No.

Listing 16.2.9.4 (Page 18 of 32)
 Listing of FVC
 (Matched Population for Piece-wise Regression Analysis on FVC)

Study: 019

| Subject ID | Visit Year | Age(yrs) at Visit | Ambulatory Status at Visit | Steroid Use | Steroid Use 24 Month | FVC (L) |
|------------|------------|-------------------|----------------------------|-------------|----------------------|---------|
| 101002 | 2015 | 15.05 | 0 | 1 | >=24 mo | 1.25 |
| | 2015 | 15.49 | 0 | 1 | >=24 mo | 1.43 |
| | 2016 | 15.95 | 0 | 1 | >=24 mo | 1.29 |
| | 2016 | 16.44 | 0 | 1 | >=24 mo | 1.48 |
| | 2017 | 16.72 | 0 | 0 | >=24 mo | 1.54 |
| 101005 | 2015 | 17.27 | 0 | 1 | >=24 mo | 1.37 |
| | 2016 | 17.79 | 0 | 1 | >=24 mo | 1.44 |
| | 2016 | 18.2 | 0 | 1 | >=24 mo | 1.21 |
| | 2017 | 18.45 | 0 | 0 | >=24 mo | 1.59 |
| 101006 | 2013 | 17.49 | 0 | 1 | >=24 mo | 2.65 |
| | 2014 | 17.94 | 0 | 1 | >=24 mo | 2.57 |
| | 2014 | 18.41 | 0 | 1 | >=24 mo | 2.55 |
| | 2015 | 18.87 | 0 | 1 | >=24 mo | 2.57 |
| | 2015 | 19.3 | 0 | 1 | >=24 mo | 2.5 |
| | 2015 | 19.78 | 0 | 1 | >=24 mo | 2.61 |
| | 2016 | 20.24 | 0 | 1 | >=24 mo | 2.66 |
| | 2016 | 20.7 | 0 | 1 | >=24 mo | 2.55 |
| | 2017 | 20.93 | 0 | 1 | >=24 mo | 2.53 |

Note: Ambulatory status: 1=ambulatory, 0=non-ambulatory. Steroid Use: 1=Yes, 0=No.

Listing 16.2.9.4 (Page 19 of 32)
 Listing of FVC
 (Matched Population for Piece-wise Regression Analysis on FVC)

Study: 019

| Subject ID | Visit Year | Age(yrs) at Visit | Ambulatory Status at Visit | Steroid Use | Steroid Use 24 Month | FVC (L) |
|------------|------------|-------------------|----------------------------|-------------|----------------------|---------|
| 101007 | 2013 | 13.27 | 0 | 1 | >=24 mo | 1.71 |
| | 2014 | 13.73 | 0 | 1 | >=24 mo | 1.59 |
| | 2014 | 14.18 | 0 | 1 | >=24 mo | 1.8 |
| | 2015 | 14.64 | 0 | 1 | >=24 mo | 1.8 |
| | 2015 | 15.09 | 0 | 1 | >=24 mo | 1.57 |
| | 2015 | 15.57 | 0 | 1 | >=24 mo | 1.62 |
| | 2016 | 15.99 | 0 | 1 | >=24 mo | 1.76 |
| | 2016 | 16.48 | 0 | 1 | >=24 mo | 1.95 |
| | 2017 | 16.72 | 0 | 0 | >=24 mo | 2.02 |
| 101008 | 2013 | 14.28 | 0 | 0 | >=24 mo | 1.42 |
| | 2013 | 14.73 | 0 | 1 | >=24 mo | 1.52 |
| | 2014 | 15.19 | 0 | 0 | >=24 mo | 1.47 |
| 102001 | 2015 | 15.12 | 0 | 1 | >=24 mo | 2.72 |
| | 2016 | 16.08 | 0 | 1 | >=24 mo | 2.78 |
| | 2017 | 16.71 | 0 | 1 | >=24 mo | 2.73 |
| 102002 | 2013 | 13.91 | 0 | 0 | >=24 mo | 1.49 |
| | 2013 | 14.36 | 0 | 1 | >=24 mo | 1.44 |
| | 2014 | 14.83 | 0 | 1 | >=24 mo | 1.84 |

Note: Ambulatory status: 1=ambulatory, 0=non-ambulatory. Steroid Use: 1=Yes, 0=No.

Listing 16.2.9.4 (Page 20 of 32)
Listing of FVC
(Matched Population for Piece-wise Regression Analysis on FVC)

Study: 019

| Subject ID | Visit Year | Age(yrs) at Visit | Ambulatory Status at Visit | Steroid Use | Steroid Use 24 Month | FVC (L) |
|------------|------------|-------------------|----------------------------|-------------|----------------------|---------|
| 102002 | 2014 | 15.27 | 0 | 1 | >=24 mo | 1.79 |
| | 2015 | 15.75 | 0 | 1 | >=24 mo | 1.71 |
| | 2015 | 16.2 | 0 | 1 | >=24 mo | 1.5 |
| | 2015 | 16.65 | 0 | 1 | >=24 mo | 1.74 |
| | 2016 | 17.13 | 0 | 1 | >=24 mo | 1.4 |
| | 2017 | 17.74 | 0 | 1 | >=24 mo | 1.62 |
| | 103001 | 2013 | 14.98 | 0 | 1 | >=24 mo |
| 2014 | | 15.44 | 0 | 1 | >=24 mo | 1.87 |
| 2014 | | 15.9 | 0 | 1 | >=24 mo | 1.65 |
| 2015 | | 16.38 | 0 | 1 | >=24 mo | 2.14 |
| 2015 | | 16.8 | 0 | 1 | >=24 mo | 2.1 |
| 2015 | | 17.27 | 0 | 1 | >=24 mo | 1.85 |
| 2016 | | 17.77 | 0 | 1 | >=24 mo | 2.19 |
| 2016 | | 18.18 | 0 | 1 | >=24 mo | 2.02 |
| 2017 | | 18.45 | 0 | 1 | >=24 mo | 2.23 |
| 103003 | 2013 | 16.46 | 0 | 1 | >=24 mo | 2.79 |
| | 2014 | 16.91 | 0 | 1 | >=24 mo | 2.96 |
| | 2014 | 17.39 | 0 | 1 | >=24 mo | 2.88 |

Note: Ambulatory status: 1=ambulatory, 0=non-ambulatory. Steroid Use: 1=Yes, 0=No.

Listing 16.2.9.4 (Page 21 of 32)
Listing of FVC
(Matched Population for Piece-wise Regression Analysis on FVC)

Study: 019

| Subject ID | Visit Year | Age(yrs) at Visit | Ambulatory Status at Visit | Steroid Use | Steroid Use 24 Month | FVC (L) |
|------------|------------|-------------------|----------------------------|-------------|----------------------|---------|
| 103003 | 2014 | 17.85 | 0 | 1 | >=24 mo | 2.79 |
| | 2015 | 18.37 | 0 | 1 | >=24 mo | 2.96 |
| | 2015 | 18.76 | 0 | 1 | >=24 mo | 2.92 |
| | 2016 | 19.22 | 0 | 1 | >=24 mo | 2.76 |
| | 2016 | 19.45 | 0 | 0 | >=24 mo | 2.77 |
| 103004 | 2016 | 16.74 | 0 | 1 | >=24 mo | 1.25 |
| | 2017 | 17.45 | 0 | 1 | >=24 mo | 1.13 |
| 103006 | 2015 | 15.82 | 0 | 0 | >=24 mo | 3.62 |
| | 2016 | 16.24 | 0 | 0 | >=24 mo | 3.58 |
| | 2016 | 16.7 | 0 | 0 | >=24 mo | 3.57 |
| | 2017 | 17.16 | 0 | 0 | >=24 mo | 3.05 |

Note: Ambulatory status: 1=ambulatory, 0=non-ambulatory. Steroid Use: 1=Yes, 0=No.

Listing 16.2.9.4 (Page 22 of 32)
Listing of FVC
(Matched Population for Piece-wise Regression Analysis on FVC)

Study: CINRG

| Subject ID | Visit Year | Age(yrs) at Visit | Ambulatory Status at Visit | Steroid Use | Steroid Use 24 Month | FVC (L) |
|------------|------------|-------------------|----------------------------|-------------|----------------------|---------|
| 042 | 2012 | 24.17 | 0 | 1 | >=24 mo | 1.42 |
| 049 | 2012 | 23.54 | 0 | 1 | >=24 mo | 1.56 |
| | 2013 | 24.55 | 0 | 1 | >=24 mo | 1.27 |
| 051 | 2013 | 23.98 | 0 | 1 | >=24 mo | 1.92 |
| | 2013 | 24.9 | 0 | 1 | >=24 mo | 1.7 |
| 055 | 2013 | 24.01 | 0 | 1 | >=24 mo | 0.38 |
| | 2013 | 24.6 | 0 | 1 | >=24 mo | 0.46 |
| 060 | 2012 | 22.76 | 0 | 1 | >=24 mo | 0.83 |
| | 2014 | 24.75 | 0 | 1 | >=24 mo | 1.17 |
| 066 | 2014 | 24.08 | 0 | 0 | >=24 mo | 0.28 |
| 067 | 2012 | 22.32 | 0 | 0 | >=24 mo | 0.31 |
| 068 | 2012 | 22.2 | 0 | 1 | >=24 mo | 1.03 |
| | 2013 | 23.71 | 0 | 1 | >=24 mo | 0.76 |
| | 2014 | 24.71 | 0 | 1 | >=24 mo | 0.79 |
| 072 | 2012 | 21.95 | 0 | 1 | >=24 mo | 0.48 |
| | 2013 | 22.9 | 0 | 1 | >=24 mo | 0.52 |
| | 2014 | 23.94 | 0 | 1 | >=24 mo | 0.5 |
| | 2015 | 24.96 | 0 | 1 | >=24 mo | 0.5 |

Note: Ambulatory status: 1=ambulatory, 0=non-ambulatory. Steroid Use: 1=Yes, 0=No.

Listing 16.2.9.4 (Page 23 of 32)
Listing of FVC
(Matched Population for Piece-wise Regression Analysis on FVC)

Study: CINRG

| Subject ID | Visit Year | Age(yrs) at Visit | Ambulatory Status at Visit | Steroid Use | Steroid Use 24 Month | FVC (L) |
|------------|------------|-------------------|----------------------------|-------------|----------------------|---------|
| 078 | 2012 | 20.78 | 0 | 1 | >=24 mo | 3.45 |
| 079 | 2012 | 20.56 | 0 | 1 | >=24 mo | 2.64 |
| 088 | 2012 | 20.48 | 0 | 1 | >=24 mo | 1.14 |
| 089 | 2013 | 21.73 | 0 | 0 | >=24 mo | 4.5 |
| | 2014 | 22.67 | 0 | 0 | >=24 mo | 3.97 |
| 090 | 2012 | 20.57 | 0 | 1 | >=24 mo | 0.33 |
| 094 | 2012 | 20.15 | 0 | 1 | >=24 mo | 2.78 |
| 096 | 2012 | 19.57 | 0 | 1 | >=24 mo | 1.74 |
| 114 | 2012 | 19.13 | 0 | 1 | >=24 mo | 1.83 |
| | 2013 | 20.14 | 0 | 1 | >=24 mo | 1.82 |
| | 2014 | 21.19 | 0 | 1 | >=24 mo | 1.79 |
| | 2016 | 23.04 | 0 | 1 | >=24 mo | 1.55 |
| 120 | 2012 | 18.19 | 0 | 1 | >=24 mo | 2.09 |
| | 2014 | 20.88 | 0 | 1 | >=24 mo | 1.36 |
| | 2016 | 22.03 | 0 | 1 | >=24 mo | 4.68 |
| 121 | 2012 | 18.81 | 0 | 0 | >=24 mo | 1.82 |
| | 2013 | 19.78 | 0 | 0 | >=24 mo | 1.39 |
| | 2014 | 20.8 | 0 | 0 | >=24 mo | 1.21 |

Note: Ambulatory status: 1=ambulatory, 0=non-ambulatory. Steroid Use: 1=Yes, 0=No.

Listing 16.2.9.4 (Page 24 of 32)
Listing of FVC
(Matched Population for Piece-wise Regression Analysis on FVC)

Study: CINRG

| Subject ID | Visit Year | Age(yrs) at Visit | Ambulatory Status at Visit | Steroid Use | Steroid Use 24 Month | FVC (L) |
|------------|------------|-------------------|----------------------------|-------------|----------------------|---------|
| 121 | 2015 | 21.68 | 0 | 0 | >=24 mo | 1.12 |
| 122 | 2012 | 18.53 | 0 | 1 | >=24 mo | 1.12 |
| | 2013 | 19.17 | 0 | 1 | >=24 mo | 1.12 |
| | 2014 | 20.6 | 0 | 1 | >=24 mo | 0.9 |
| | 2015 | 21.58 | 0 | 1 | >=24 mo | 0.75 |
| 123 | 2012 | 18.35 | 0 | 1 | >=24 mo | 0.95 |
| | 2015 | 21.08 | 0 | 1 | >=24 mo | 1.09 |
| | 2016 | 22.27 | 0 | 1 | >=24 mo | 1.17 |
| 126 | 2012 | 18.32 | 0 | 1 | >=24 mo | 1.75 |
| | 2013 | 19.78 | 0 | 1 | >=24 mo | 1.41 |
| | 2015 | 21.02 | 0 | 1 | >=24 mo | 1.24 |
| | 2016 | 22.04 | 0 | 1 | >=24 mo | 1.11 |
| 128 | 2012 | 18.19 | 0 | 1 | >=24 mo | 2.43 |
| 131 | 2012 | 17.91 | 0 | 1 | >=24 mo | 3.69 |
| 133 | 2012 | 17.91 | 0 | 1 | >=24 mo | 1.8 |
| | 2013 | 18.88 | 0 | 1 | >=24 mo | 1.42 |
| | 2014 | 19.95 | 0 | 1 | >=24 mo | 1.51 |
| 134 | 2013 | 18.47 | 0 | 1 | >=24 mo | 1.69 |

Note: Ambulatory status: 1=ambulatory, 0=non-ambulatory. Steroid Use: 1=Yes, 0=No.

Listing 16.2.9.4 (Page 25 of 32)
Listing of FVC
(Matched Population for Piece-wise Regression Analysis on FVC)

Study: CINRG

| Subject ID | Visit Year | Age(yrs) at Visit | Ambulatory Status at Visit | Steroid Use | Steroid Use 24 Month | FVC (L) |
|------------|------------|-------------------|----------------------------|-------------|----------------------|---------|
| 134 | 2013 | 18.78 | 0 | 1 | >=24 mo | 1.98 |
| 135 | 2012 | 17.45 | 0 | 1 | >=24 mo | 3.43 |
| 136 | 2012 | 18.04 | 0 | 1 | >=24 mo | 1.13 |
| 140 | 2013 | 18.26 | 0 | 1 | >=24 mo | 0.68 |
| 144 | 2012 | 17.24 | 0 | 1 | >=24 mo | 0.9 |
| 147 | 2012 | 17.33 | 0 | 1 | >=24 mo | 2.77 |
| 148 | 2012 | 16.86 | 0 | 1 | >=24 mo | 1.63 |
| | 2013 | 17.98 | 0 | 1 | >=24 mo | 1.29 |
| | 2014 | 19.04 | 0 | 1 | >=24 mo | 1.35 |
| | 2016 | 20.93 | 0 | 1 | >=24 mo | 1.16 |
| 152 | 2013 | 18.02 | 0 | 1 | >=24 mo | 2.11 |
| | 2014 | 19.03 | 0 | 1 | >=24 mo | 2.01 |
| | 2016 | 20.97 | 0 | 1 | >=24 mo | 1.84 |
| 157 | 2012 | 16.48 | 0 | 0 | >=24 mo | 0.47 |
| | 2013 | 17.33 | 0 | 0 | >=24 mo | 0.3 |
| | 2015 | 19.57 | 0 | 0 | >=24 mo | 0.29 |
| 159 | 2012 | 16.81 | 0 | 0 | >=24 mo | 1.38 |
| | 2013 | 17.83 | 0 | 0 | >=24 mo | 1.13 |

Note: Ambulatory status: 1=ambulatory, 0=non-ambulatory. Steroid Use: 1=Yes, 0=No.

Listing 16.2.9.4 (Page 26 of 32)
 Listing of FVC
 (Matched Population for Piece-wise Regression Analysis on FVC)

Study: CINRG

| Subject ID | Visit Year | Age(yrs) at Visit | Ambulatory Status at Visit | Steroid Use | Steroid Use 24 Month | FVC (L) |
|------------|------------|-------------------|----------------------------|-------------|----------------------|---------|
| 159 | 2014 | 18.85 | 0 | 0 | >=24 mo | 0.83 |
| | 2015 | 19.82 | 0 | 0 | >=24 mo | 0.63 |
| 160 | 2012 | 16.33 | 0 | 0 | >=24 mo | 2.35 |
| 161 | 2012 | 16.56 | 0 | 1 | >=24 mo | 2 |
| | 2014 | 17.91 | 0 | 1 | >=24 mo | 2.1 |
| | 2015 | 18.95 | 0 | 1 | >=24 mo | 2.22 |
| | 2016 | 19.98 | 0 | 1 | >=24 mo | 1.78 |
| 169 | 2012 | 15.52 | 0 | 0 | >=24 mo | 1.59 |
| 170 | 2013 | 17.18 | 0 | 1 | >=24 mo | 0.78 |
| | 2014 | 18.04 | 0 | 1 | >=24 mo | 0.79 |
| 179 | 2012 | 15.55 | 0 | 0 | >=24 mo | 1.48 |
| | 2013 | 16.56 | 0 | 0 | >=24 mo | 1.49 |
| 181 | 2012 | 15.52 | 0 | 1 | >=24 mo | 1.55 |
| | 2014 | 18.08 | 0 | 1 | >=24 mo | 0.96 |
| | 2015 | 19.08 | 0 | 1 | >=24 mo | 0.96 |
| 182 | 2012 | 15.62 | 0 | 1 | >=24 mo | 2.53 |
| 183 | 2013 | 17.1 | 0 | 0 | >=24 mo | 1.16 |
| 184 | 2012 | 15.26 | 0 | 1 | >=24 mo | 1.65 |

Note: Ambulatory status: 1=ambulatory, 0=non-ambulatory. Steroid Use: 1=Yes, 0=No.

Listing 16.2.9.4 (Page 27 of 32)
 Listing of FVC
 (Matched Population for Piece-wise Regression Analysis on FVC)

Study: CINRG

| Subject ID | Visit Year | Age(yrs) at Visit | Ambulatory Status at Visit | Steroid Use | Steroid Use 24 Month | FVC (L) |
|------------|------------|-------------------|----------------------------|-------------|----------------------|---------|
| 184 | 2013 | 16.45 | 0 | 1 | >=24 mo | 1.42 |
| | 2014 | 17.38 | 0 | 1 | >=24 mo | 1.31 |
| 187 | 2012 | 15.28 | 0 | 0 | >=24 mo | 1.74 |
| | 2013 | 16.35 | 0 | 0 | >=24 mo | 1.36 |
| 189 | 2012 | 15.23 | 0 | 1 | >=24 mo | 1.38 |
| 190 | 2013 | 15.98 | 0 | 0 | >=24 mo | 0.69 |
| | 2013 | 16.73 | 0 | 0 | >=24 mo | 1.3 |
| 191 | 2012 | 15.25 | 0 | 1 | >=24 mo | 1.77 |
| 193 | 2012 | 15.43 | 0 | 1 | >=24 mo | 1.23 |
| 194 | 2012 | 14.98 | 0 | 1 | >=24 mo | 1.69 |
| | 2013 | 15.93 | 0 | 1 | >=24 mo | 1.82 |
| | 2014 | 16.88 | 0 | 1 | >=24 mo | 1.68 |
| 195 | 2012 | 15.46 | 0 | 0 | >=24 mo | 3.11 |
| 198 | 2012 | 15 | 0 | 0 | >=24 mo | 2.24 |
| | 2013 | 15.56 | 0 | 0 | >=24 mo | 1.41 |
| | 2014 | 16.66 | 0 | 0 | >=24 mo | 1.75 |
| | 2015 | 17.7 | 0 | 0 | >=24 mo | 1.81 |
| 199 | 2016 | 18.43 | 0 | 1 | >=24 mo | 1.5 |

Note: Ambulatory status: 1=ambulatory, 0=non-ambulatory. Steroid Use: 1=Yes, 0=No.

Listing 16.2.9.4 (Page 28 of 32)
Listing of FVC
(Matched Population for Piece-wise Regression Analysis on FVC)

Study: CINRG

| Subject ID | Visit Year | Age(yrs) at Visit | Ambulatory Status at Visit | Steroid Use | Steroid Use 24 Month | FVC (L) |
|------------|------------|-------------------|----------------------------|-------------|----------------------|---------|
| 202 | 2013 | 15.68 | 0 | 1 | >=24 mo | 2.47 |
| 203 | 2012 | 14.02 | 0 | 1 | >=24 mo | 1.72 |
| | 2013 | 15.14 | 0 | 1 | >=24 mo | 2.19 |
| | 2014 | 16.19 | 0 | 1 | >=24 mo | 2.11 |
| | 2016 | 18.08 | 0 | 1 | >=24 mo | 2.22 |
| 204 | 2013 | 15.2 | 0 | 1 | >=24 mo | 1.53 |
| | 2014 | 16.21 | 0 | 1 | >=24 mo | 1.82 |
| 205 | 2012 | 14.33 | 0 | 1 | >=24 mo | 2.66 |
| | 2013 | 15.14 | 0 | 1 | >=24 mo | 2.66 |
| | 2014 | 16.48 | 0 | 1 | >=24 mo | 2.23 |
| | 2015 | 17.11 | 0 | 1 | >=24 mo | 2.18 |
| 206 | 2013 | 15.37 | 0 | 1 | >=24 mo | 1.2 |
| | 2014 | 15.9 | 0 | 1 | >=24 mo | 0.99 |
| | 2015 | 17.33 | 0 | 1 | >=24 mo | 0.82 |
| 207 | 2013 | 14.98 | 0 | 1 | >=24 mo | 1.48 |
| | 2013 | 15.33 | 0 | 1 | >=24 mo | 1.41 |
| | 2014 | 16.32 | 0 | 1 | >=24 mo | 1.32 |
| 210 | 2013 | 15.05 | 0 | 1 | >=24 mo | 2.54 |

Note: Ambulatory status: 1=ambulatory, 0=non-ambulatory. Steroid Use: 1=Yes, 0=No.

Listing 16.2.9.4 (Page 29 of 32)
Listing of FVC
(Matched Population for Piece-wise Regression Analysis on FVC)

Study: CINRG

| Subject ID | Visit Year | Age(yrs) at Visit | Ambulatory Status at Visit | Steroid Use | Steroid Use 24 Month | FVC (L) |
|------------|------------|-------------------|----------------------------|-------------|----------------------|---------|
| 210 | 2014 | 15.96 | 0 | 1 | >=24 mo | 2.5 |
| | 2015 | 17.08 | 0 | 1 | >=24 mo | 2.88 |
| | 2016 | 18.16 | 0 | 1 | >=24 mo | 2.78 |
| 217 | 2014 | 16.02 | 0 | 1 | >=24 mo | 2.4 |
| | 2015 | 16.96 | 0 | 1 | >=24 mo | 2.17 |
| 219 | 2013 | 14.72 | 0 | 0 | >=24 mo | 1.87 |
| | 2014 | 15.89 | 0 | 0 | >=24 mo | 1.22 |
| 222 | 2012 | 13.94 | 0 | 1 | >=24 mo | 2.17 |
| 225 | 2012 | 13.59 | 0 | 1 | >=24 mo | 1.41 |
| 227 | 2013 | 15.14 | 0 | 1 | >=24 mo | 1.15 |
| | 2014 | 16.13 | 0 | 1 | >=24 mo | 1.41 |
| 228 | 2012 | 13.57 | 0 | 0 | >=24 mo | 1.59 |
| | 2013 | 14.42 | 0 | 0 | >=24 mo | 1.35 |
| | 2014 | 15.61 | 0 | 0 | >=24 mo | 1.21 |
| | 2015 | 16.65 | 0 | 0 | >=24 mo | 1.03 |
| 231 | 2012 | 14.08 | 0 | 1 | >=24 mo | 1.18 |
| | 2013 | 15.11 | 0 | 1 | >=24 mo | 0.73 |
| | 2014 | 16.13 | 0 | 1 | >=24 mo | 0.62 |

Note: Ambulatory status: 1=ambulatory, 0=non-ambulatory. Steroid Use: 1=Yes, 0=No.

Listing 16.2.9.4 (Page 30 of 32)
Listing of FVC
(Matched Population for Piece-wise Regression Analysis on FVC)

Study: CINRG

| Subject ID | Visit Year | Age(yrs) at Visit | Ambulatory Status at Visit | Steroid Use | Steroid Use 24 Month | FVC (L) |
|------------|------------|-------------------|----------------------------|-------------|----------------------|---------|
| 233 | 2013 | 14.83 | 0 | 0 | >=24 mo | 1.85 |
| | 2015 | 16.89 | 0 | 0 | >=24 mo | 1.56 |
| 234 | 2012 | 13.5 | 0 | 1 | >=24 mo | 1.6 |
| | 2013 | 14.53 | 0 | 1 | >=24 mo | 1.88 |
| | 2014 | 15.57 | 0 | 1 | >=24 mo | 1.89 |
| | 2015 | 16.58 | 0 | 1 | >=24 mo | 1.7 |
| 240 | 2013 | 14.41 | 0 | 1 | >=24 mo | 0.88 |
| | 2015 | 15.48 | 0 | 1 | >=24 mo | 0.77 |
| 241 | 2013 | 14.22 | 0 | 1 | >=24 mo | 1.6 |
| 243 | 2012 | 13.23 | 0 | 0 | >=24 mo | 2.41 |
| | 2014 | 15.23 | 0 | 0 | >=24 mo | 2.65 |
| 252 | 2012 | 12.13 | 0 | 1 | >=24 mo | 1.28 |
| 258 | 2016 | 15.88 | 0 | 1 | >=24 mo | 1.52 |
| 261 | 2012 | 11.7 | 0 | 1 | >=24 mo | 2.02 |
| | 2014 | 13.83 | 0 | 1 | >=24 mo | 1.9 |
| 266 | 2012 | 12.08 | 0 | 1 | >=24 mo | 1.83 |
| | 2013 | 13.16 | 0 | 1 | >=24 mo | 2.01 |
| | 2014 | 14.16 | 0 | 1 | >=24 mo | 1.93 |

Note: Ambulatory status: 1=ambulatory, 0=non-ambulatory. Steroid Use: 1=Yes, 0=No.

Listing 16.2.9.4 (Page 31 of 32)
Listing of FVC
(Matched Population for Piece-wise Regression Analysis on FVC)

Study: CINRG

| Subject ID | Visit Year | Age(yrs) at Visit | Ambulatory Status at Visit | Steroid Use | Steroid Use 24 Month | FVC (L) |
|------------|------------|-------------------|----------------------------|-------------|----------------------|---------|
| 272 | 2012 | 12 | 0 | 1 | >=24 mo | 1.26 |
| | 2013 | 13.01 | 0 | 1 | >=24 mo | 0.98 |
| | 2014 | 14.03 | 0 | 1 | >=24 mo | 0.85 |
| | 2015 | 15 | 0 | 1 | >=24 mo | 0.63 |
| 274 | 2012 | 11.84 | 0 | 1 | >=24 mo | 1.6 |
| | 2013 | 12.96 | 0 | 0 | >=24 mo | 1.57 |
| 275 | 2013 | 12.48 | 0 | 0 | >=24 mo | 2.1 |
| | 2014 | 13.63 | 0 | 0 | >=24 mo | 2.21 |
| 283 | 2012 | 11.5 | 0 | 0 | >=24 mo | 2 |
| | 2014 | 13.33 | 0 | 0 | >=24 mo | 1.4 |
| 284 | 2012 | 10.88 | 0 | 0 | >=24 mo | 2.29 |
| | 2013 | 11.77 | 0 | 1 | >=24 mo | 2.46 |
| 285 | 2012 | 11.21 | 0 | 1 | >=24 mo | 1.53 |
| | 2014 | 13.29 | 0 | 1 | >=24 mo | 1.76 |
| | 2015 | 14.39 | 0 | 1 | >=24 mo | 1.75 |
| 293 | 2015 | 13.98 | 0 | 1 | >=24 mo | 2.47 |
| 297 | 2014 | 12.48 | 0 | 1 | >=24 mo | 1.7 |
| | 2015 | 13.42 | 0 | 1 | >=24 mo | 1.75 |

Note: Ambulatory status: 1=ambulatory, 0=non-ambulatory. Steroid Use: 1=Yes, 0=No.

Listing 16.2.9.4 (Page 32 of 32)
 Listing of FVC
 (Matched Population for Piece-wise Regression Analysis on FVC)

Study: CINRG

| Subject ID | Visit Year | Age(yrs) at Visit | Ambulatory Status at Visit | Steroid Use | Steroid Use 24 Month | FVC (L) |
|------------|------------|-------------------|----------------------------|-------------|----------------------|---------|
| 300 | 2015 | 12.76 | 0 | 1 | >=24 mo | 1.86 |
| | 2016 | 13.75 | 0 | 1 | >=24 mo | 2.01 |
| 316 | 2012 | 9.44 | 0 | 1 | >=24 mo | 1.92 |
| | 2013 | 10.35 | 0 | 1 | >=24 mo | 1.75 |
| | 2014 | 11.35 | 0 | 1 | >=24 mo | 2.08 |
| | 2015 | 12.54 | 0 | 1 | >=24 mo | 2.17 |
| 317 | 2012 | 9.53 | 0 | 1 | >=24 mo | 1.79 |
| | 2014 | 10.65 | 0 | 0 | >=24 mo | 2 |
| | 2015 | 11.63 | 0 | 1 | >=24 mo | 2.27 |
| | 2015 | 12.52 | 0 | 1 | >=24 mo | 2.21 |
| 321 | 2016 | 12.54 | 0 | 1 | >=24 mo | 1.19 |
| 322 | 2015 | 11.62 | 0 | 1 | >=24 mo | 2.07 |
| | 2016 | 12.51 | 0 | 1 | >=24 mo | 2.62 |
| 325 | 2013 | 9.28 | 0 | 0 | >=24 mo | 2.08 |
| | 2014 | 10.18 | 0 | 0 | >=24 mo | 2.09 |
| | 2015 | 11.21 | 0 | 0 | >=24 mo | 2.38 |
| | 2016 | 12.31 | 0 | 0 | >=24 mo | 2.34 |
| 342 | 2016 | 10.76 | 0 | 1 | >=24 mo | 1.96 |

Note: Ambulatory status: 1=ambulatory, 0=non-ambulatory. Steroid Use: 1=Yes, 0=No.

Listing 16.2.9.5a (Page 1 of 2)
 Age at Loss of Ambulation for Study 019
 (Matched Population for Age at Loss of Ambulation)

Study: 019

| Subject ID | Age(yrs)/ Race | Loss of Ambulation (Event/Censor) | Age at Loss of Ambulation or Censor(Years) | Loss of Ambulation (Event/Censor) Based on AE | Age at Loss of Ambulation or Censor(Years) Based on AE |
|------------|-----------------------|--------------------------------------|--|---|---|
| 001013 | 15.21/White/Caucasian | Yes (Event) | 16.8 | Yes (Event) | 16.8 |
| 003003 | 15.37/Asian | Yes (Event) | 16.2 | Yes (Event) | 16.2 |
| 011001 | 14.91/White/Caucasian | Yes (Event) | 16.3 | Yes (Event) | 16.3 |
| 011003 | 15.91/White/Caucasian | Yes (Event) | 16.9 | Yes (Event) | 16.9 |
| 012005 | 13.63/White/Caucasian | Yes (Event) | 15.5 | Yes (Event) | 15.5 |
| 021002 | 15.93/White/Caucasian | No (Censor) | 20.8 | No (Censor) | 20.8 |
| 021003 | 13.59/White/Caucasian | No (Censor) | 18.4 | No (Censor) | 18.4 |
| 021004 | 12.67/White/Caucasian | Yes (Event) | 16.4 | Yes (Event) | 16.4 |
| 021005 | 12.62/White/Caucasian | Yes (Event) | 17.4 | Yes (Event) | 17.4 |
| 031002 | 11.32/White/Caucasian | Yes (Event) | 15 | Yes (Event) | 15 |
| 031004 | 13.46/White/Caucasian | No (Censor) | 18.3 | No (Censor) | 18.3 |
| 033001 | 10.11/White/Caucasian | No (Censor) | 14.7 | No (Censor) | 14.7 |
| 034001 | 13.48/White/Caucasian | Yes (Event) | 13.9 | Yes (Event) | 13.9 |
| 034002 | 14/Asian | Yes (Event) | 14.9 | Yes (Event) | 14.9 |
| 041001 | 15.06/White/Caucasian | No (Censor) | 19.7 | No (Censor) | 19.7 |
| 041003 | 12.99/White/Caucasian | No (Censor) | 17.6 | No (Censor) | 17.6 |
| 051004 | 12.12/White/Caucasian | No (Censor) | 12.1 | No (Censor) | 12.1 |
| 061002 | 12.02/White/Caucasian | Yes (Event) | 12.5 | Yes (Event) | 12.5 |
| 062002 | 12.04/White/Caucasian | Yes (Event) | 15.5 | Yes (Event) | 15.5 |
| 062004 | 12.77/White/Caucasian | Yes (Event) | 15.5 | Yes (Event) | 15.5 |
| 062006 | 13.77/White/Caucasian | No (Censor) | 18.1 | No (Censor) | 18.1 |
| 062007 | 11.32/Unknown | No (Censor) | 15.5 | No (Censor) | 15.5 |
| 063001 | 11/White/Caucasian | Yes (Event) | 15 | Yes (Event) | 15 |
| 063003 | 9.96/White/Caucasian | No (Censor) | 14.6 | No (Censor) | 14.6 |
| 063004 | 10.25/White/Caucasian | No (Censor) | 14.8 | No (Censor) | 14.8 |

Listing 16.2.9.5a (Page 2 of 2)
 Age at Loss of Ambulation for Study 019
 (Matched Population for Age at Loss of Ambulation)

Study: 019

| Subject ID | Age(yrs) / Race | Loss of Ambulation (Event/Censor) | Age at Loss of Ambulation or Censor(Years) | Loss of Ambulation (Event/Censor) Based on AE | Age at Loss of Ambulation or Censor(Years) Based on AE |
|------------|-----------------------|--------------------------------------|--|---|---|
| 063005 | 9.43/White/Caucasian | Yes (Event) | 12.5 | Yes (Event) | 12.5 |
| 071002 | 10.41/White/Caucasian | Yes (Event) | 13.1 | Yes (Event) | 13.1 |
| 081001 | 12.89/White/Caucasian | Yes (Event) | 14.3 | Yes (Event) | 14.3 |
| 082001 | 11.43/White/Caucasian | No (Censor) | 12.3 | No (Censor) | 12.3 |
| 082002 | 11.16/White/Caucasian | No (Censor) | 15.4 | No (Censor) | 15.4 |
| 091001 | 9.29/White/Caucasian | Yes (Event) | 12.8 | Yes (Event) | 12.8 |
| 091003 | 9.14/White/Caucasian | Yes (Event) | 10.9 | Yes (Event) | 10.9 |
| 091005 | 12.29/White/Caucasian | No (Censor) | 16.7 | No (Censor) | 16.7 |
| 092001 | 13.17/White/Caucasian | Yes (Event) | 15 | Yes (Event) | 15 |
| 101001 | 11.96/White/Caucasian | No (Censor) | 15.9 | No (Censor) | 15.9 |
| 101002 | 12.75/White/Caucasian | Yes (Event) | 14.1 | Yes (Event) | 14.1 |
| 101003 | 12.06/White/Caucasian | No (Censor) | 16 | No (Censor) | 16 |
| 101005 | 14.52/White/Caucasian | Yes (Event) | 17 | Yes (Event) | 17 |
| 101006 | 17.02/White/Caucasian | Yes (Event) | 17.5 | Yes (Event) | 17.5 |
| 101009 | 13.67/White/Caucasian | No (Censor) | 17.6 | No (Censor) | 17.6 |
| 101011 | 9.23/White/Caucasian | No (Censor) | 13.2 | No (Censor) | 13.2 |
| 102001 | 12.82/Asian | Yes (Event) | 15.1 | Yes (Event) | 15.1 |
| 103004 | 13.98/White/Caucasian | Yes (Event) | 16.4 | Yes (Event) | 16.4 |
| 103006 | 13.48/White/Caucasian | Yes (Event) | 15.3 | Yes (Event) | 15.3 |

Listing 16.2.9.5b (Page 1 of 2)
Age at Loss of Ambulation for Study CINRG
(Matched Population for Age at Loss of Ambulation)

Study: CINRG

| Subject ID | Age(yrs) / Race | Loss of Ambulation (Event/Censor) | Age at Loss of Ambulation or Censor(Years) |
|------------|-----------------------|--------------------------------------|--|
| 075 | 16.7/White/Caucasian | No (Censor) | 21.8 |
| 080 | 16.75/White/Caucasian | Yes (Event) | 17.5 |
| 085 | 15.71/White/Caucasian | No (Censor) | 19.8 |
| 089 | 15.67/White/Caucasian | Yes (Event) | 18.7 |
| 094 | 15.08/White/Caucasian | Yes (Event) | 17.1 |
| 104 | 13.55/White/Caucasian | Yes (Event) | 14.6 |
| 107 | 13.44/White/Caucasian | Yes (Event) | 15.4 |
| 110 | 13.96/White/Caucasian | No (Censor) | 16.1 |
| 114 | 14.14/White/Caucasian | Yes (Event) | 19.1 |
| 120 | 13.18/White/Caucasian | Yes (Event) | 15.2 |
| 122 | 13.48/Asian | Yes (Event) | 16.4 |
| 127 | 13.82/White/Caucasian | No (Censor) | 21.8 |
| 128 | 13.18/White/Caucasian | Yes (Event) | 18.2 |
| 130 | 13.17/White/Caucasian | No (Censor) | 17.2 |
| 135 | 12.31/White/Caucasian | Yes (Event) | 15.3 |
| 141 | 12.09/White/Caucasian | No (Censor) | 20.8 |
| 143 | 13.82/White/Caucasian | No (Censor) | 20.9 |
| 144 | 12.23/White/Caucasian | Yes (Event) | 13.2 |
| 145 | 12.43/White/Caucasian | No (Censor) | 20.5 |
| 146 | 12.34/White/Caucasian | No (Censor) | 20.5 |
| 147 | 12.26/White/Caucasian | Yes (Event) | 16.3 |
| 148 | 11.83/White/Caucasian | Yes (Event) | 13.9 |
| 149 | 11.21/White/Caucasian | Yes (Event) | 14.2 |
| 151 | 11.68/White/Caucasian | No (Censor) | 15.7 |
| 154 | 12.07/White/Caucasian | No (Censor) | 12.1 |

Listing 16.2.9.5b (Page 2 of 2)
Age at Loss of Ambulation for Study CINRG
(Matched Population for Age at Loss of Ambulation)

Study: CINRG

| Subject ID | Age(yrs) / Race | Loss of Ambulation (Event/Censor) | Age at Loss of Ambulation or Censor(Years) |
|------------|------------------------------|--------------------------------------|--|
| 156 | 11.78/White/Caucasian | No (Censor) | 12.4 |
| 161 | 11.43/White/Caucasian | Yes (Event) | 14.7 |
| 165 | 11.06/White/Caucasian | Yes (Event) | 11.3 |
| 167 | 10.42/Black/African American | No (Censor) | 15.7 |
| 178 | 10.11/Asian | Yes (Event) | 13.2 |
| 181 | 10.52/White/Caucasian | Yes (Event) | 15.5 |
| 186 | 10.33/White/Caucasian | No (Censor) | 11.9 |
| 188 | 11.07/Pacific Islander | No (Censor) | 13.1 |
| 190 | 9.73/White/Caucasian | Yes (Event) | 10.7 |
| 191 | 10.18/Other | Yes (Event) | 12.1 |
| 193 | 10.38/White/Caucasian | Yes (Event) | 14.6 |
| 194 | 9.8/White/Caucasian | Yes (Event) | 15 |
| 195 | 11.47/Other | Yes (Event) | 15.5 |
| 196 | 9.97/White/Caucasian | No (Censor) | 18 |
| 197 | 9.13/Asian | Yes (Event) | 11.1 |
| 199 | 9.28/White/Caucasian | Yes (Event) | 18.4 |
| 205 | 9.38/White/Caucasian | Yes (Event) | 10.8 |
| 206 | 9.86/White/Caucasian | Yes (Event) | 12.9 |
| 211 | 9.28/Asian | Yes (Event) | 11.3 |
| 228 | 9.34/White/Caucasian | Yes (Event) | 10.3 |
| 249 | 9.19/Asian | No (Censor) | 15.1 |

Listing 16.2.9.6 (Page 1 of 5)
 Age at FVC <1 Liter
 (Matched Population for Age at FVC < 1 Liter Analysis)

Study: 019

| Subject ID | Age (yrs) / Race | FVC <1 Liter (Event/Censor) | Age at FVC <1 Liter or Censor (Years) [1] |
|------------|-----------------------|--------------------------------|--|
| 011002 | 14.06/White/Caucasian | No (Censor) | 14 |
| 011004 | 15.49/White/Caucasian | No (Censor) | 19.2 |
| 011005 | 13.41/White/Caucasian | No (Censor) | 17.1 |
| 011006 | 13.07/White/Caucasian | Yes (Event) | 16.3 |
| 012001 | 15.73/White/Caucasian | No (Censor) | 19.4 |
| 021001 | 14.77/White/Caucasian | No (Censor) | 19.6 |
| 031003 | 11.78/White/Caucasian | Yes (Event) | 13.7 |
| 041004 | 13.59/White/Caucasian | No (Censor) | 18.2 |
| 041005 | 12.69/White/Caucasian | No (Censor) | 17.3 |
| 042001 | 11.03/White/Caucasian | No (Censor) | 15.6 |
| 042002 | 12.65/White/Caucasian | No (Censor) | 17.3 |
| 043001 | 12.29/White/Caucasian | No (Censor) | 14.6 |
| 043002 | 14.36/Asian | No (Censor) | 19 |
| 051003 | 15.87/White/Caucasian | No (Censor) | 20 |
| 052002 | 15.2/White/Caucasian | No (Censor) | 17 |
| 052003 | 12.98/White/Caucasian | No (Censor) | 17.6 |
| 052006 | 11.48/White/Caucasian | No (Censor) | 15.5 |
| 061001 | 12/White/Caucasian | No (Censor) | 16.6 |
| 062001 | 14.28/White/Caucasian | No (Censor) | 18.4 |
| 062003 | 13.6/White/Caucasian | No (Censor) | 14.2 |
| 063002 | 13.21/Other | No (Censor) | 13.7 |
| 063006 | 9.41/White/Caucasian | No (Censor) | 10.4 |
| 071001 | 12.63/White/Caucasian | No (Censor) | 17.3 |
| 081002 | 10.22/White/Caucasian | No (Censor) | 14.5 |
| 082003 | 14.05/White/Caucasian | No (Censor) | 18.3 |

Listing 16.2.9.6 (Page 2 of 5)
Age at FVC <1 Liter
(Matched Population for Age at FVC < 1 Liter Analysis)

Study: 019

| Subject ID | Age (yrs) / Race | FVC <1 Liter (Event/Censor) | Age at FVC <1 Liter or Censor (Years) [1] |
|------------|-----------------------|--------------------------------|--|
| 082004 | 14.33/White/Caucasian | No (Censor) | 14.9 |
| 091004 | 15.27/White/Caucasian | Yes (Event) | 18 |
| 092002 | 15.73/White/Caucasian | No (Censor) | 18 |
| 092003 | 14.28/Other | No (Censor) | 14.7 |
| 101004 | 10.86/White/Caucasian | Yes (Event) | 15.2 |
| 101007 | 12.8/White/Caucasian | No (Censor) | 16.7 |
| 101008 | 14.28/White/Caucasian | No (Censor) | 15.2 |
| 102002 | 13.91/White/Caucasian | No (Censor) | 17.7 |
| 102003 | 12.33/White/Caucasian | No (Censor) | 14.2 |
| 103001 | 14.98/White/Caucasian | No (Censor) | 18.5 |
| 103003 | 16.46/White/Caucasian | No (Censor) | 19.5 |
| 103007 | 13.6/White/Caucasian | No (Censor) | 17 |

Listing 16.2.9.6 (Page 3 of 5)
Age at FVC <1 Liter
(Matched Population for Age at FVC < 1 Liter Analysis)

Study: CINRG

| Subject ID | Age (yrs) / Race | FVC <1 Liter (Event/Censor) | Age at FVC <1 Liter or Censor (Years) [1] |
|------------|------------------------------|--------------------------------|--|
| 052 | 17.83/White/Caucasian | Yes (Event) | 18.8 |
| 068 | 17.17/White/Caucasian | Yes (Event) | 19.2 |
| 071 | 16.18/White/Caucasian | Yes (Event) | 17 |
| 072 | 17.88/White/Caucasian | Yes (Event) | 18.5 |
| 074 | 17.8/Other | Yes (Event) | 18.3 |
| 077 | 16.93/White/Caucasian | No (Censor) | 16.9 |
| 079 | 15.62/Asian | No (Censor) | 20.6 |
| 081 | 15.7/Other | No (Censor) | 22.8 |
| 082 | 16.26/White/Caucasian | Yes (Event) | 19.3 |
| 084 | 15.8/White/Caucasian | Yes (Event) | 24 |
| 088 | 16.37/White/Caucasian | No (Censor) | 21.4 |
| 091 | 14.58/Asian | Yes (Event) | 22.6 |
| 093 | 15.61/Other | Yes (Event) | 17.8 |
| 095 | 15.25/White/Caucasian | Yes (Event) | 22.3 |
| 096 | 14.45/White/Caucasian | No (Censor) | 19.6 |
| 099 | 14.82/Asian | No (Censor) | 18.9 |
| 102 | 14.53/Black/African American | No (Censor) | 20.1 |
| 106 | 13.67/White/Caucasian | Yes (Event) | 15 |
| 109 | 13.38/White/Caucasian | No (Censor) | 17.5 |
| 111 | 15.25/White/Caucasian | No (Censor) | 18.2 |
| 115 | 12.78/Other | No (Censor) | 12.8 |
| 117 | 13.68/White/Caucasian | No (Censor) | 17.9 |
| 118 | 12.68/Other | Yes (Event) | 22 |
| 121 | 13.74/Other | No (Censor) | 21.7 |
| 123 | 13.34/White/Caucasian | Yes (Event) | 18.4 |

Listing 16.2.9.6 (Page 4 of 5)
 Age at FVC <1 Liter
 (Matched Population for Age at FVC < 1 Liter Analysis)

Study: CINRG

| Subject ID | Age (yrs) / Race | FVC <1 Liter (Event/Censor) | Age at FVC <1 Liter or Censor (Years) [1] |
|------------|-----------------------|--------------------------------|--|
| 125 | 12.78/White/Caucasian | Yes (Event) | 14.4 |
| 126 | 14.33/White/Caucasian | No (Censor) | 22 |
| 132 | 12.91/White/Caucasian | Yes (Event) | 19.2 |
| 134 | 11.74/White/Caucasian | No (Censor) | 18.8 |
| 136 | 12.95/Asian | No (Censor) | 18 |
| 137 | 12.02/Other | No (Censor) | 16.1 |
| 138 | 14.11/Asian | No (Censor) | 16.2 |
| 139 | 14.11/Asian | No (Censor) | 16.1 |
| 140 | 12.65/White/Caucasian | Yes (Event) | 12.9 |
| 150 | 12.11/White/Caucasian | Yes (Event) | 13.3 |
| 157 | 12.51/White/Caucasian | Yes (Event) | 16.5 |
| 159 | 12.82/White/Caucasian | Yes (Event) | 18.9 |
| 160 | 11.32/White/Caucasian | No (Censor) | 16.3 |
| 162 | 11.36/Asian | Yes (Event) | 12.4 |
| 163 | 11.91/Other | No (Censor) | 16.9 |
| 164 | 10.45/Asian | No (Censor) | 14.5 |
| 169 | 11.48/Other | No (Censor) | 16 |
| 170 | 12.17/White/Caucasian | Yes (Event) | 15.3 |
| 174 | 11.68/Asian | No (Censor) | 13.7 |
| 175 | 12.36/Asian | No (Censor) | 16.9 |
| 179 | 10.58/White/Caucasian | No (Censor) | 17.7 |
| 182 | 10.53/White/Caucasian | No (Censor) | 15.6 |
| 184 | 10.34/White/Caucasian | No (Censor) | 17.4 |
| 187 | 10.25/White/Caucasian | No (Censor) | 16.4 |
| 208 | 10.03/Asian | No (Censor) | 14.2 |

Listing 16.2.9.6 (Page 5 of 5)
Age at FVC <1 Liter
(Matched Population for Age at FVC < 1 Liter Analysis)

Study: CINRG

| Subject ID | Age (yrs) / Race | FVC <1 Liter (Event/Censor) | Age at FVC <1 Liter or Censor (Years) [1] |
|------------|----------------------|--------------------------------|--|
| 233 | 9.83/White/Caucasian | No (Censor) | 16.9 |
| 245 | 9.05/Asian | Yes (Event) | 14.4 |