## CH50 (Week 1-2)

| Patient | Treatment | Visit Date time | D1 Pre-dose | D1 Post-dose | Week1 D2-D7 Pre-plasmapheresis | Week 1 D2-D7 Post-plasmapheresis | Week 1 D2-D7 1hr post-intervention | Week2 D8-D14 Pre-plasmapheresis | Week2 D8-D14 Post-plasmapheresis | Week2 D8-D14 1hr post-intervention |
| --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- |
| ######## | xx | YYYY-MM-DD | ##.## | ##.## | ##.## | ##.## | ##.## | ##.## | ##.## | ##.## |
| ######## | xx | YYYY-MM-DD | ##.## | ##.## | ##.## | ##.## | ##.## | ##.## | ##.## | ##.## |
| ######## | xx | YYYY-MM-DD | ##.## | ##.## | ##.## | ##.## | ##.## | ##.## | ##.## | ##.## |
| ######## | xx | YYYY-MM-DD | ##.## | ##.## | ##.## | ##.## | ##.## | ##.## | ##.## | ##.## |
| ######## | xx | YYYY-MM-DD | ##.## | ##.## | ##.## | ##.## | ##.## | ##.## | ##.## | ##.## |
| LLOQ - lower limit of quantification. Detection limit xx Eq/mL | | | | | | | | | | |

## CH50 (Week 4-49)

| Visit | Cohort A | | Cohort B | |
| --- | --- | --- | --- | --- |
|  | BIV020+SOC | SOC | BIV020+SOC | SOC |
|  | N = ## | N = ## | N = ## | N = ## |
| CH50 D1 Pre-dose |  |  |  |  |
| N | ## | ## | ## | ## |
| Mean (SD) | ##.## (##.##) | ##.## (##.##) | ##.## (##.##) | ##.## (##.##) |
| Median | ##.## | ##.## | ##.## | ##.## |
| Q1:Q3 | ##.##: ##.## | ##.##: ##.## | ##.##: ##.## | ##.##: ##.## |
| Min: Max | ##.##: ##.## | ##.##: ##.## | ##.##: ##.## | ##.##: ##.## |
| CH50 Week 4 |  |  |  |  |
| N | ## | ## | ## | ## |
| Mean (SD) | ##.## (##.##) | ##.## (##.##) | ##.## (##.##) | ##.## (##.##) |
| Median | ##.## | ##.## | ##.## | ##.## |
| Q1:Q3 | ##.##: ##.## | ##.##: ##.## | ##.##: ##.## | ##.##: ##.## |
| Min: Max | ##.##: ##.## | ##.##: ##.## | ##.##: ##.## | ##.##: ##.## |
| CH50 Week 13 |  |  |  |  |
| N | ## | ## | ## | ## |
| Mean (SD) | ##.## (##.##) | ##.## (##.##) | ##.## (##.##) | ##.## (##.##) |
| Median | ##.## | ##.## | ##.## | ##.## |
| Q1:Q3 | ##.##: ##.## | ##.##: ##.## | ##.##: ##.## | ##.##: ##.## |
| Min: Max | ##.##: ##.## | ##.##: ##.## | ##.##: ##.## | ##.##: ##.## |
| CH50 Week 25 |  |  |  |  |
| N | ## | ## | ## | ## |
| Mean (SD) | ##.## (##.##) | ##.## (##.##) | ##.## (##.##) | ##.## (##.##) |
| Median | ##.## | ##.## | ##.## | ##.## |
| Q1:Q3 | ##.##: ##.## | ##.##: ##.## | ##.##: ##.## | ##.##: ##.## |
| Min: Max | ##.##: ##.## | ##.##: ##.## | ##.##: ##.## | ##.##: ##.## |
| CH50 Week 37 |  |  |  |  |
| N | ## | ## | ## | ## |
| Mean (SD) | ##.## (##.##) | ##.## (##.##) | ##.## (##.##) | ##.## (##.##) |
| Median | ##.## | ##.## | ##.## | ##.## |
| Q1:Q3 | ##.##: ##.## | ##.##: ##.## | ##.##: ##.## | ##.##: ##.## |
| Min: Max | ##.##: ##.## | ##.##: ##.## | ##.##: ##.## | ##.##: ##.## |
| CH50 Week 49 |  |  |  |  |
| N | ## | ## | ## | ## |
| Mean (SD) | ##.## (##.##) | ##.## (##.##) | ##.## (##.##) | ##.## (##.##) |
| Median | ##.## | ##.## | ##.## | ##.## |
| Q1:Q3 | ##.##: ##.## | ##.##: ##.## | ##.##: ##.## | ##.##: ##.## |
| Min: Max | ##.##: ##.## | ##.##: ##.## | ##.##: ##.## | ##.##: ##.## |
| CH50 End of Study |  |  |  |  |
| N | ## | ## | ## | ## |
| Mean (SD) | ##.## (##.##) | ##.## (##.##) | ##.## (##.##) | ##.## (##.##) |
| Median | ##.## | ##.## | ##.## | ##.## |
| Q1:Q3 | ##.##: ##.## | ##.##: ##.## | ##.##: ##.## | ##.##: ##.## |
| Min: Max | ##.##: ##.## | ##.##: ##.## | ##.##: ##.## | ##.##: ##.## |
| For the plasmapheresis performed in W1-W2 (BIVV020 + SOC arm only), complement classical pathway activity samples (CP and CH50) were collected before (within 1 hour) and after each plasmapheresis session (within 1 hour after PP; or specifically ≤1 h before BIVV020 IV supplemental dose administration) and at 1 h after BIVV020 supplemental dose start of infusion (±10 min). ; LLOQ – lower limit of quantification | | | | |

## CH50 Change from baseline cohort A (Week 1-2)

| Visits | BIV020+SOC | SOC |
| --- | --- | --- |
| Change from baseline to Week 1 D2-D7 Postdose |  |  |
| N | ## | ## |
| Mean (SD) | ##.## (##.##) | ##.## (##.##) |
| Median | ##.## | ##.## |
| Q1:Q3 | ##.##: ##.## | ##.##: ##.## |
| Min: Max | ##.##: ##.## | ##.##: ##.## |
| N under LLOQ | ## (##.##%) | ## (##.##%) |
| Change from baseline to Week 1 D2-D7 Pre-plasmapheresis |  |  |
| N | ## | ## |
| Mean (SD) | ##.## (##.##) | ##.## (##.##) |
| Median | ##.## | ##.## |
| Q1:Q3 | ##.##: ##.## | ##.##: ##.## |
| Min: Max | ##.##: ##.## | ##.##: ##.## |
| N under LLOQ | ## (##.##%) | ## (##.##%) |
| Change from baseline to Week 1 D2-D7 1hr Post-plasmapheresis |  |  |
| N | ## | ## |
| Mean (SD) | ##.## (##.##) | ##.## (##.##) |
| Median | ##.## | ##.## |
| Q1:Q3 | ##.##: ##.## | ##.##: ##.## |
| Min: Max | ##.##: ##.## | ##.##: ##.## |
| N under LLOQ | ## (##.##%) | ## (##.##%) |
| Change from baseline to Week 1 D2-D7 1hr Post-intervention |  |  |
| N | ## | ## |
| Mean (SD) | ##.## (##.##) | ##.## (##.##) |
| Median | ##.## | ##.## |
| Q1:Q3 | ##.##: ##.## | ##.##: ##.## |
| Min: Max | ##.##: ##.## | ##.##: ##.## |
| N under LLOQ | ## (##.##%) | ## (##.##%) |
| Change from baseline to Week 2 D8-D14 Pre-plasmapheresis |  |  |
| N | ## | ## |
| Mean (SD) | ##.## (##.##) | ##.## (##.##) |
| Median | ##.## | ##.## |
| Q1:Q3 | ##.##: ##.## | ##.##: ##.## |
| Min: Max | ##.##: ##.## | ##.##: ##.## |
| N under LLOQ | ## (##.##%) | ## (##.##%) |
| Change from baseline to Week 2 D8-D14 1hr Post-plasmapheresis |  |  |
| N | ## | ## |
| Mean (SD) | ##.## (##.##) | ##.## (##.##) |
| Median | ##.## | ##.## |
| Q1:Q3 | ##.##: ##.## | ##.##: ##.## |
| Min: Max | ##.##: ##.## | ##.##: ##.## |
| N under LLOQ | ## (##.##%) | ## (##.##%) |
| Change from baseline to Week 2 D8-D14 1hr Post-intervention |  |  |
| N | ## | ## |
| Mean (SD) | ##.## (##.##) | ##.## (##.##) |
| Median | ##.## | ##.## |
| Q1:Q3 | ##.##: ##.## | ##.##: ##.## |
| Min: Max | ##.##: ##.## | ##.##: ##.## |
| N under LLOQ | ## (##.##%) | ## (##.##%) |
| Values below detection limit were imputed with d/√ 2; due the small sample size the test between two groups are not provided. | | |

## CH50 Change from baseline cohort A (Week 4-49)

| Visits | BIV020+SOC | SOC |
| --- | --- | --- |
| Change from baseline to Week 4 |  |  |
| N | ## | ## |
| Mean (SD) | ##.## (##.##) | ##.## (##.##) |
| Median | ##.## | ##.## |
| Q1:Q3 | ##.##: ##.## | ##.##: ##.## |
| Min: Max | ##.##: ##.## | ##.##: ##.## |
| Change from baseline to Week 13 |  |  |
| N | ## | ## |
| Mean (SD) | ##.## (##.##) | ##.## (##.##) |
| Median | ##.## | ##.## |
| Q1:Q3 | ##.##: ##.## | ##.##: ##.## |
| Min: Max | ##.##: ##.## | ##.##: ##.## |
| Change from baseline to Week 25 |  |  |
| N | ## | ## |
| Mean (SD) | ##.## (##.##) | ##.## (##.##) |
| Median | ##.## | ##.## |
| Q1:Q3 | ##.##: ##.## | ##.##: ##.## |
| Min: Max | ##.##: ##.## | ##.##: ##.## |
| Change from baseline to Week 37 |  |  |
| N | ## | ## |
| Mean (SD) | ##.## (##.##) | ##.## (##.##) |
| Median | ##.## | ##.## |
| Q1:Q3 | ##.##: ##.## | ##.##: ##.## |
| Min: Max | ##.##: ##.## | ##.##: ##.## |
| Change from baseline to Week 49 |  |  |
| N | ## | ## |
| Mean (SD) | ##.## (##.##) | ##.## (##.##) |
| Median | ##.## | ##.## |
| Q1:Q3 | ##.##: ##.## | ##.##: ##.## |
| Min: Max | ##.##: ##.## | ##.##: ##.## |

## CH50 Change from baseline Cohort B(Week 1-2)

|  | BIV020+SOC | SOC |
| --- | --- | --- |
| Change from baseline to Week 1 D2-D7 Post-plasmapheresis |  |  |
| N | ## | ## |
| Mean (SD) | ##.## (##.##) | ##.## (##.##) |
| LS Mean (95% CI) | ##.## (##.##) | ##.## (##.##) |
|  |  |  |
| LS Mean difference BIV020+SOC vs. SOC | ##.## (##.##) | - |
| 95% CI | ##.##: ##.## | - |
| p-value | #.### | - |
| Change from baseline to Week 1 D2-D7 1hr post-intervention |  |  |
| N | ## | ## |
| Mean (SD) | ##.## (##.##) | ##.## (##.##) |
| LS Mean (95% CI) | ##.## (##.##) | ##.## (##.##) |
|  |  |  |
| LS Mean difference BIV020+SOC vs. SOC | ##.## (##.##) | - |
| 95% CI | ##.##: ##.## | - |
| p-value | #.### | - |
| Change from baseline to Week 2 D8-D14 Pre-plasmapheresis |  |  |
| N | ## | ## |
| Mean (SD) | ##.## (##.##) | ##.## (##.##) |
| LS Mean (95% CI) | ##.## (##.##) | ##.## (##.##) |
|  |  |  |
| LS Mean difference BIV020+SOC vs. SOC | ##.## (##.##) | - |
| 95% CI | ##.##: ##.## | - |
| p-value | #.### | - |
| Change from baseline to Week 2 D8-D14 Post-plasmapheresis |  |  |
| N | ## | ## |
| Mean (SD) | ##.## (##.##) | ##.## (##.##) |
| LS Mean (95% CI) | ##.## (##.##) | ##.## (##.##) |
|  |  |  |
| LS Mean difference BIV020+SOC vs. SOC | ##.## (##.##) | - |
| 95% CI | ##.##: ##.## | - |
| p-value | #.### | - |
| Change from baseline to Week 2 D8-D14 1hr post-intervention |  |  |
| N | ## | ## |
| Mean (SD) | ##.## (##.##) | ##.## (##.##) |
| LS Mean (95% CI) | ##.## (##.##) | ##.## (##.##) |
|  |  |  |
| LS Mean difference BIV020+SOC vs. SOC | ##.## (##.##) | - |
| 95% CI | ##.##: ##.## | - |
| p-value | #.### | - |
| Based on an ANCOVA model after adjusting baseline value. Values below detection limit were imputed with d/√; ANCOVA = Analysis of Covariance, CI = Confidence Interval, LS = Least Squares, SD = Standard Deviation | | |

## CH50 Change from baseline (Week 4-49)

| Visits | BIV020+SOC | SOC |
| --- | --- | --- |
| Change from baseline to Week 4 |  |  |
| N | ## | ## |
| Mean (SD) | ##.## (##.##) | ##.## (##.##) |
| Median | ##.## | ##.## |
| Q1:Q3 | ##.##: ##.## | ##.##: ##.## |
| Min: Max | ##.##: ##.## | ##.##: ##.## |
| Change from baseline to Week 13 |  |  |
| N | ## | ## |
| Mean (SD) | ##.## (##.##) | ##.## (##.##) |
| Median | ##.## | ##.## |
| Q1:Q3 | ##.##: ##.## | ##.##: ##.## |
| Min: Max | ##.##: ##.## | ##.##: ##.## |
| Change from baseline to Week 25 |  |  |
| N | ## | ## |
| Mean (SD) | ##.## (##.##) | ##.## (##.##) |
| Median | ##.## | ##.## |
| Q1:Q3 | ##.##: ##.## | ##.##: ##.## |
| Min: Max | ##.##: ##.## | ##.##: ##.## |
| Change from baseline to Week 37 |  |  |
| N | ## | ## |
| Mean (SD) | ##.## (##.##) | ##.## (##.##) |
| Median | ##.## | ##.## |
| Q1:Q3 | ##.##: ##.## | ##.##: ##.## |
| Min: Max | ##.##: ##.## | ##.##: ##.## |
| Change from baseline to Week 49 |  |  |
| N | ## | ## |
| Mean (SD) | ##.## (##.##) | ##.## (##.##) |
| Median | ##.## | ##.## |
| Q1:Q3 | ##.##: ##.## | ##.##: ##.## |
| Min: Max | ##.##: ##.## | ##.##: ##.## |