KINIKSA

*Every Second Counts!™*

# Mavrilimumab Phase 2 GCA Data

*November 2020*

# Forward Looking Statements

This presentation (together with any other statements or information that we may make in connection herewith) contains forward-looking statements within the meaning of the Private Securities Litigation Reform Act of 1995 with respect to Kiniksa Pharmaceuticals, Ltd. (and its consolidated subsidiaries, collectively, unless context otherwise requires, "Kiniksa," "we," "us" or "our"). In some cases, you can identify forward looking statements by terms such as "may," "will," "should," "expect," "plan," "anticipate," "could," "intend," "goal," "design," "target," "project," "contemplate," "believe," "estimate," "predict," "potential" or "continue" or the negative of these terms or other similar expressions, although not all forward-looking statements contain these identifying words. All statements contained in this presentation that do not relate to matters of historical fact should be considered forward-looking statements, including without limitation, statements regarding our strategy; potential acquisitions and collaborations; potential value drivers; potential indications; potential market opportunities and competitive position; on going, planned and potential clinical trials and other studies; timing and potential impact of clinical data; regulatory and other submissions, applications and approvals; commercial strategy and pre-commercial activities; expected run rate for our cash, cash equivalents and short-term investments; expected funding of our operating plan; and capital allocation.

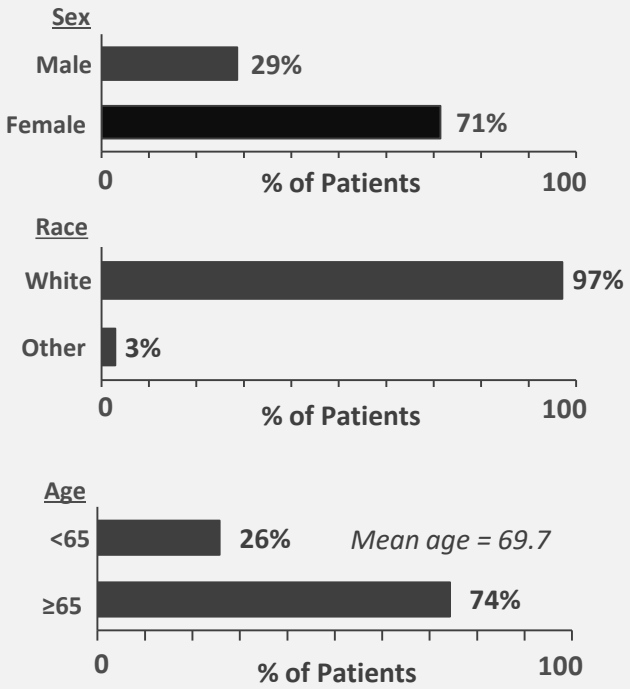
These statements involve known and unknown risks, uncertainties, and other important factors that may cause our actual results, performance or achievements to be materially different from those expressed or implied by the forward-looking statements, including without limitation potential delays or difficulties with our clinical trials; potential inability to demonstrate safety or efficacy or otherwise producing negative, inconclusive or uncompetitive results; potential for changes in final data from preliminary or interim data; potential inability to replicate in later clinical trials positive results from earlier trials and studies; our reliance on third parties for manufacturing and conducting clinical trials, research and other studies; potential changes in our strategy, operating plan and funding requirements; drug substance and/or drug product shortages; substantial new or existing competition; potential impact of the COVID-19 pandemic, and measures taken in response to the pandemic, on our business and operations as well as the business and operations of our manufacturers, CROs upon whom we rely to conduct our clinical trials, and other third parties with whom we conduct business or otherwise engage, including the FDA and other regulatory authorities; and our ability to attract and retain qualified personnel. These and the other important factors are discussed under the caption "Risk Factors" in our Quarterly Report on Form 10-Q filed with the Securities and Exchange Commission (the "SEC") on November 5, 2020 and other filings subsequently filed with the SEC. These forward-looking statements reflect various assumptions of Kiniksa's management that may or may not prove to be correct. No forward-looking statement is a guarantee of future results, performance, or achievements, and one should avoid placing undue reliance on such statements. Except as otherwise indicated, this presentation speaks as of the date of this presentation. We undertake no obligation to update any forward-looking statements, whether as a result of new information, future events or otherwise.

This presentation also contains estimates, projections, and/or other information regarding our industry, our business and the markets for certain of our product candidates, including data regarding the estimated size of those markets, and the incidence and prevalence of certain medical conditions. Unless otherwise expressly stated, we obtained this industry, business, market and other data from reports, research surveys, clinical trials, studies and similar data prepared by market research firms and other third parties, from industry, medical and general publications, and from government data and similar sources. Information that is based on estimates, forecasts, projections, market research, or similar methodologies is inherently subject to uncertainties and actual events or circumstances may differ materially from events and circumstances reflected in this information.

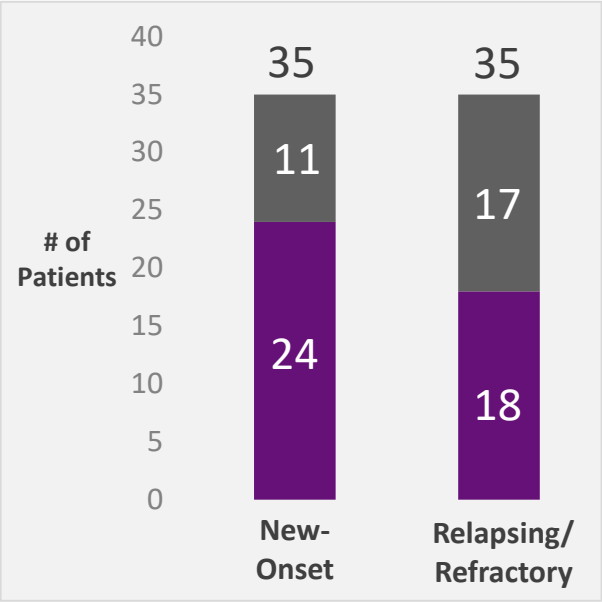
# Baseline Demographics and Clinical Characteristics

## Mavrilimumab Phase 2 Giant Cell Arteritis Data

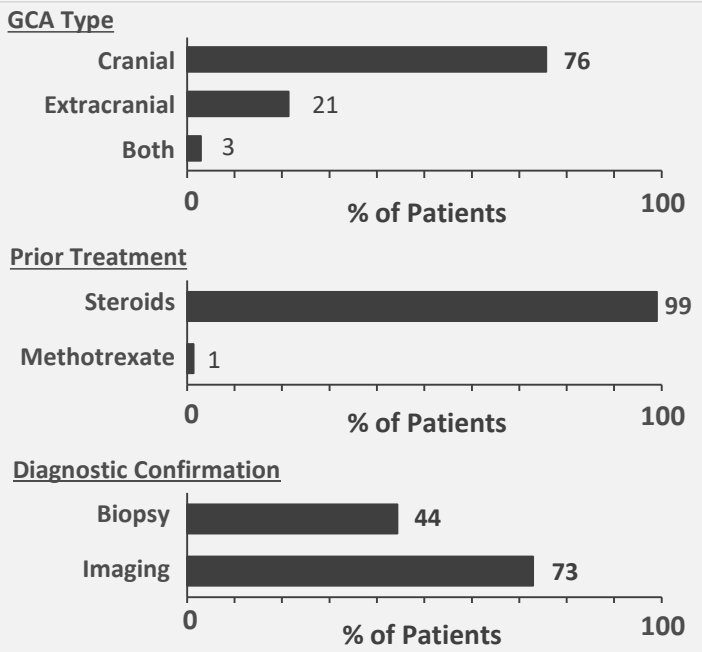
### Baseline Demographics (n=70)




### Randomization Strata




### Baseline Disease Characteristics (n=70)



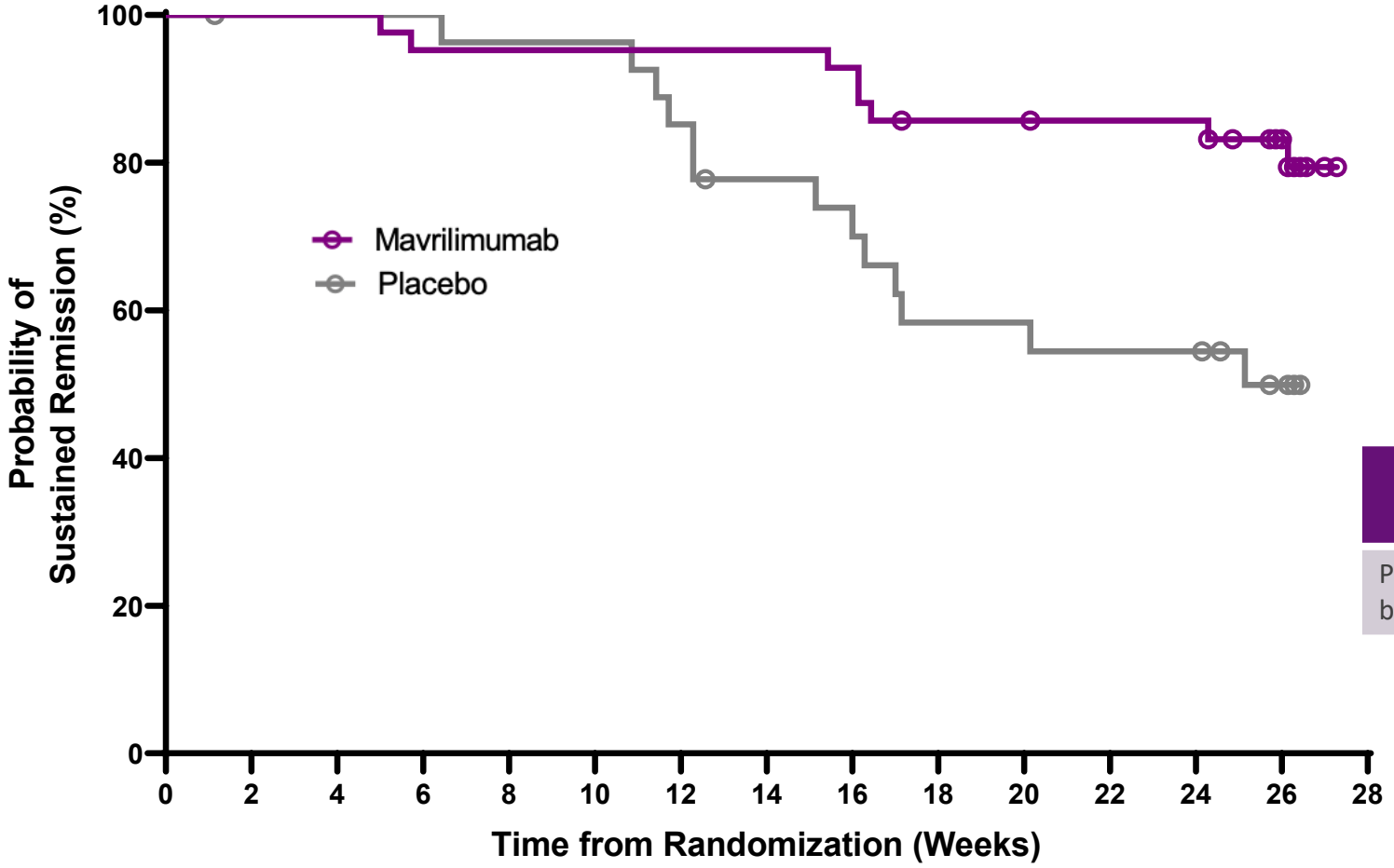

Mean time since diagnosis = 1.1 months (new-onset)
Mean time since diagnosis = 16.2 months (relapsing/refractory)

Mean eligibility ESR = 56.2 mm/hr

Mean eligibility CRP = 4.27 mg/dL

# Primary Efficacy Endpoint: Time-to-First Adjudicated GCA Flare by Week 26

## Mavrilimumab Phase 2 Giant Cell Arteritis Data



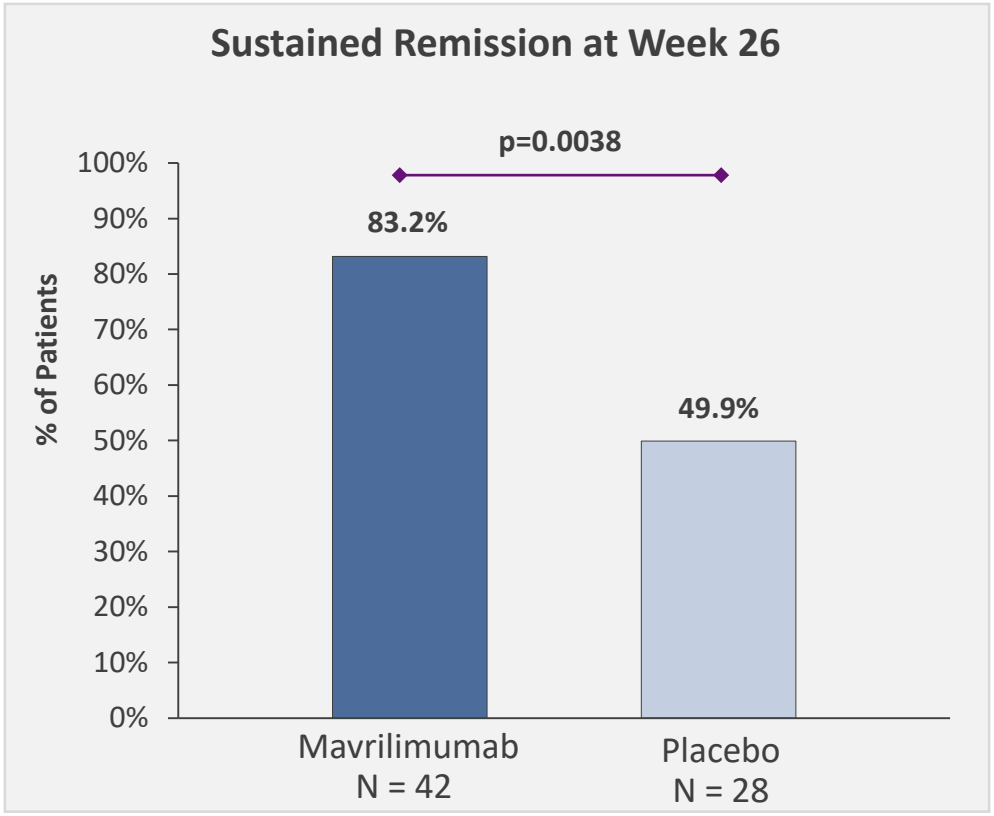

**HR = 0.38**
**Risk Reduction = 62%**
**p=0.0263**

| | Mavrilimumab (N=42) | Placebo (N=28) |
|---|---|---|
| Patients with Flare by Week 26, n (%) | 8 (19) | 13 (46.4) |

*Median time-to-flare by Week 26 could not be estimated in mavrilimumab recipients due to the low number of flares in the mavrilimumab treatment arm. The median time-to-flare for placebo recipients was 25.1 weeks. There was a 62% lower risk of flare in mavrilimumab recipients compared to placebo recipients*

# Secondary Efficacy Endpoint: Sustained Remission at Week 26

## Mavrilimumab Phase 2 Giant Cell Arteritis Data



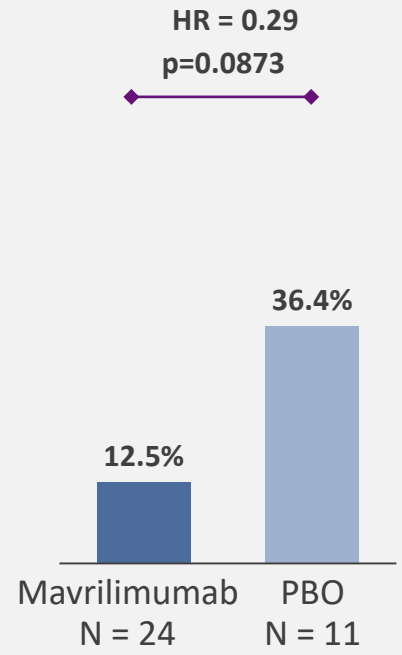

*The sustained remission rate at Week 26 was 33.3 percentage points higher in mavrilimumab recipients (83.2%) compared to placebo recipients (49.9%) (p=0.0038).*

# Consistent Trend of Efficacy Across the New Onset and Relapsing/Refractory Cohorts

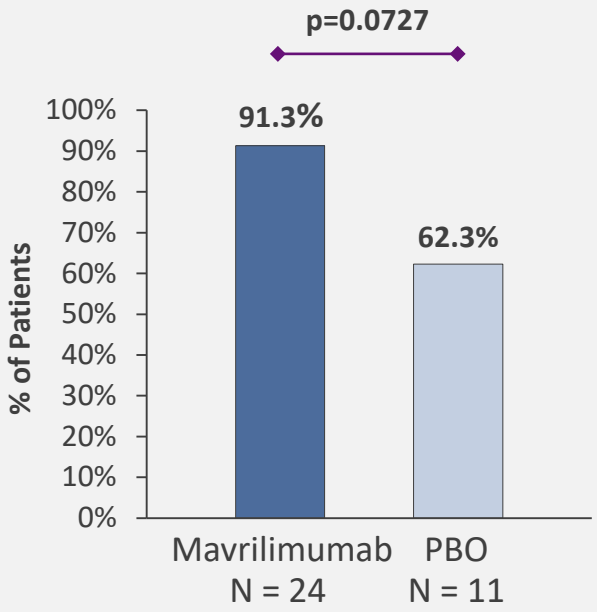
Mavrilimumab Phase 2 Giant Cell Arteritis Data

## New-Onset GCA

### Patients With Flare by Week 26




### Sustained Remission at Week 26

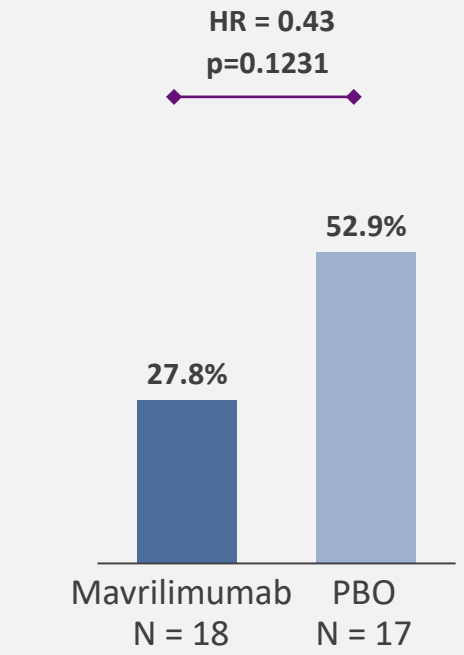



*There was a 71% lower risk of flare in mavrilimumab recipients compared to placebo recipients (Hazard Ratio = 0.29, p=0.0873).*

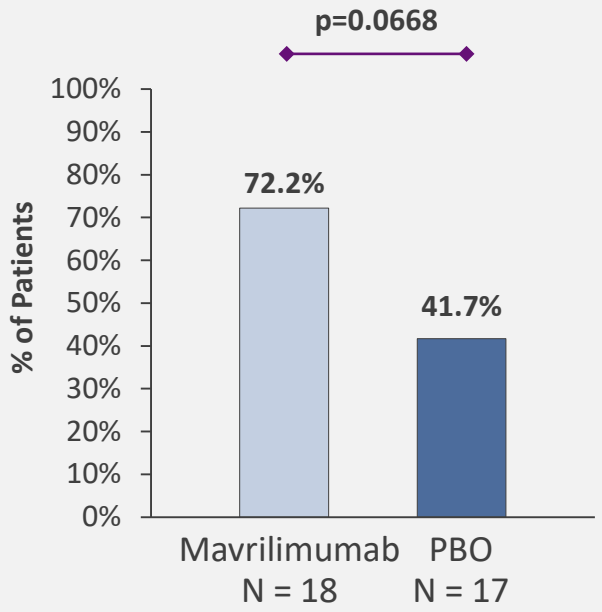
*The sustained remission rate at Week 26 was 28.9 percentage points higher in mavrilimumab recipients (91.3%) compared to placebo recipients (62.3%) (p=0.0727).*

## Relapsing/Refractory GCA

### Patients With Flare by Week 26




### Sustained Remission at Week 26




***Nominal p values**

*There was a 57% lower risk of flare in mavrilimumab recipients compared to placebo recipients (Hazard Ratio = 0.43, p=0.1231).*

*The sustained remission rate at Week 26 was 30.6 percentage points higher in mavrilimumab recipients (72.2%) compared to placebo recipients (41.7%) (p=0.0668).*

# Time to Flare and Sustained Remission at Week 26

## Mavrilimumab Phase 2 Giant Cell Arteritis Data

| Time to Flare by Week 26 and Sustained Remission at Week 26 - Total mITT Population | Mavrilimumab 150 mg (N=42) | Placebo (N=28) |
|---|---|---|
| Number of Subjects with Flare, n (%) | 8 (19.0) | 13 (46.4) |
| **Primary Efficacy Endpoint: Time to Flare (weeks) by Week 26 [1]** | | |
| Median, 95% CI | NE (NE, NE) | 25.1 (16.0, NE) |
| Hazard Ratio (Mavrilimumab vs Placebo), 95% CI [2] | 0.38 (0.15, 0.92) | |
| **P-value [3]** | 0.0263 | |
| **Secondary Efficacy Endpoint: Sustained Remission at Week 26 (%), 95% CI [4]** | 83.2 (67.9, 91.6) | 49.9 (29.6, 67.3) |
| Difference in Proportions (95% CI) [5] | 33.3 (10.7, 55.8) | |
| **P-value [5]** | 0.0038 | |

| Time to Flare by Week 26 and Sustained Remission at Week 26 by Randomization Strata | New-onset | | Relapsing/Refractory | |
|---|---|---|---|---|
| | Mavrilimumab 150 mg (N=24) | Placebo (N=11) | Mavrilimumab 150 mg (N=18) | Placebo (N=17) |
| Number of Subjects with Flare, n (%) | 3 (12.5) | 4 (36.4) | 5 (27.8) | 9 (52.9) |
| **Primary Endpoint: Time to Flare (weeks) by Week 26 [1]** | | | | |
| Median, 95% CI | NE (NE, NE) | NE (11.7, NE) | NE (16.4, NE) | 22.6 (16.0, NE) |
| Hazard Ratio (Mavrilimumab vs Placebo), 95% CI [6] | 0.29 (0.06, 1.31) | | 0.43 (0.14, 1.30) | |
| P-value [7] [8] | 0.0873 | | 0.1231 | |
| **Secondary Endpoint: Sustained Remission at Week 26 (%) , 95% CI [4]** | 91.3 (69.3, 97.7) | 62.3 (27.7, 84.0) | 72.2 (45.6, 87.4) | 41.7 (17.4, 64.5) |
| Difference in Proportions (95% CI) [5] | 28.9 (-2.7, 60.5) | | 30.6 (-2.1, 63.2) | |
| P-value [5][8] | 0.0727 | | 0.0668 | |

NE = Not estimable.

[1] Kaplan-Meier method used to estimate the survival functions for each treatment arm.

[2] Calculated based on a Cox proportional-hazards model with treatment as covariate and stratified by randomization strata.

[3] Comparison of KPL-301 and placebo with respect to time to flare calculated by using a log-rank test and stratified by randomization strata.

[4] Kaplan-Meier Survival Estimates with standard error and 95% CI for each arm.

[5] Two-sided p-value and 95% CI for the difference in sustained remission between two arms using normal approximation. Placebo arm is the reference.

[6] Calculated based on a Cox proportional-hazards model with treatment as covariate.

[7] Comparison of KPL-301 and placebo with respect to time to flare calculated by using a log-rank test.

[8] Subgroup analyses were not powered for significance; nominal p values reported.

# Summary of Adverse Events

## Mavrilimumab Phase 2 Giant Cell Arteritis Data

| | Mavrilimumab 150mg (N=42) n (%) | Placebo (N=28) n (%) |
|---|---|---|
| Treatment Emergent Adverse Events | 33 (78.6) | 25 (89.3) |
| By Maximum Severity [1] | | |
| Mild | 18 (42.9) | 13 (46.4) |
| Moderate | 14 (33.3) | 11 (39.3) |
| Severe | 1 (2.4) | 1 (3.6) |
| Related to Mavrilimumab or Placebo [2] | 10 (23.8) | 7 (25.0) |
| Related to Prednisone [2] | 11 (26.2) | 11 (39.3) |
| Serious Treatment Emergent Adverse Events | 2 (4.8) | 3 (10.7) |
| Related to Mavrilimumab or Placebo [2] | 0 | 0 |
| Related to Prednisone [2] | 0 | 0 |
| Non-serious Treatment Emergent Adverse Events | 33 (78.6) | 25 (89.3) |
| Treatment Emergent Adverse Events Resulting in Death | 0 | 0 |
| Treatment Emergent Adverse Events Leading to Dose Interruption | 1 (2.4) | 2 (7.1) |
| Treatment Emergent Adverse Events Leading to Withdrawal of Treatment | 1 (2.4) | 1 (3.6) |
| Treatment Emergent Adverse Events of Special Interest | 0 | 1 (3.6) |

*There were no drug-related serious adverse events, and the rates of drug-related treatment-emergent adverse events between mavrilimumab recipients and placebo recipients were similar*

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