

Every Second Counts![™]

RHAPSODY Phase 3 Results

November 2020

Forward Looking Statements

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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.



Summary of Pivotal Phase 3 Trial of Rilonacept in Recurrent Pericarditis



- Prespecified primary and all major secondary efficacy endpoints were highly statistically significant.
- The primary efficacy endpoint of median time-to-first adjudicated pericarditis recurrence in the randomized withdrawal period was highly statistically significant.
 - Median [95% CI] time to pericarditis recurrence for rilonacept recipients in the randomized withdrawal period could not be estimated due to the low number of recurrences in the rilonacept treatment arm. The median time-to-recurrence for placebo recipients was 8.6 [4.0-11.7] weeks (Hazard Ratio = 0.04, p<0.0001).
 - Rilonacept recipients experienced a 96% reduction in risk of recurrent pericarditis events.
- All major secondary efficacy endpoints in the randomized withdrawal period were also highly statistically significant.
 - 81% of rilonacept recipients maintained clinical response at Week 16 of the randomized withdrawal period, compared to 20% of placebo recipients (p=0.0002). Consistent results were observed at Week 8 and Week 24 and were also highly statistically significant (p<0.0001 and p=0.0022, respectively).
 - The proportion of rilonacept recipients with absent or minimal pericarditis symptoms at Week 16 of the randomized withdrawal period was 81% compared to 25% for placebo recipients (p=0.0006). Consistent results were observed at Week 8 and Week 24 and were also highly statistically significant (p<0.0001 and p=0.0002, respectively).
 - Rilonacept recipients experienced no or minimal pain for 95% of trial days through Week 16 compared to 47% of trial days for placebo recipients (p<0.0001). Consistent results were observed at Weeks 8 and 24 and were also highly significant (p<0.0001 and p<0.0001, respectively).
- Patients are continuing to receive open label rilonacept in the Long-Term Extension.
- Rilonacept was well-tolerated, with a safety profile consistent with the existing ARCALYST[®] label.
- Based on the Phase 3 RHAPSODY data, Kiniksa plans to submit an sBLA with the FDA later this year.



3

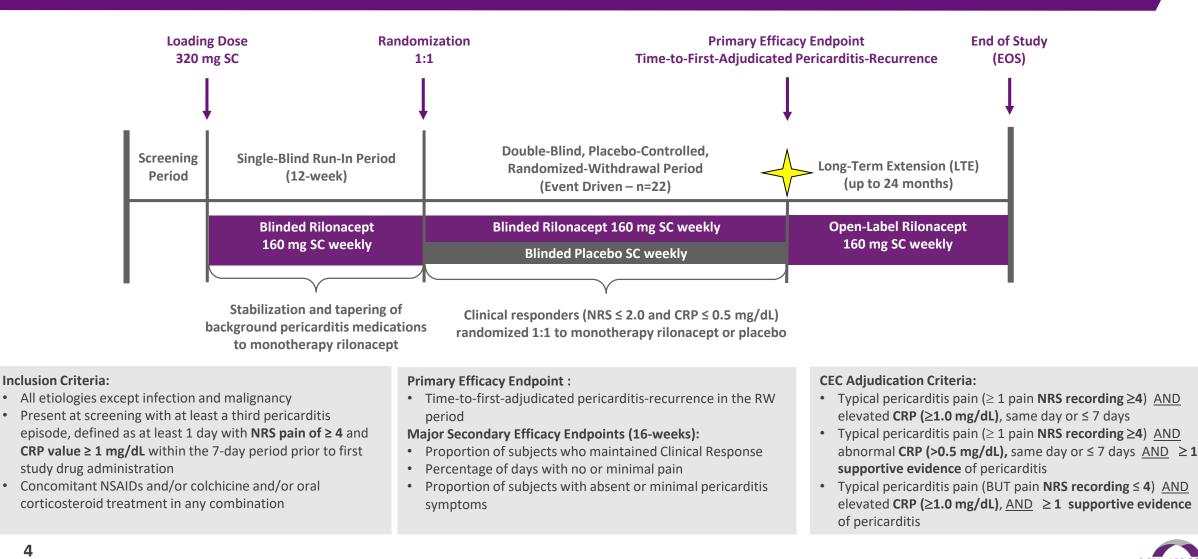
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Design of Pivotal Phase 3 Trial of Rilonacept in Recurrent Pericarditis



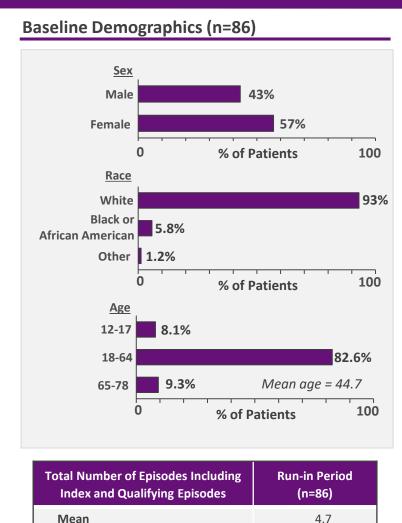




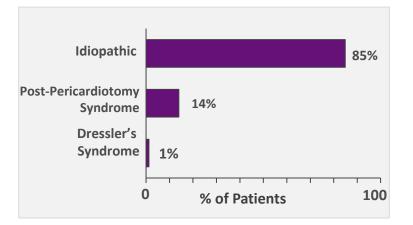
Baseline Demographics and Clinical Characteristics

Pivotal Phase 3 Rilonacept Data

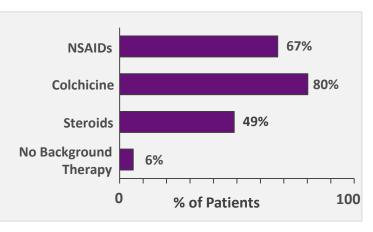




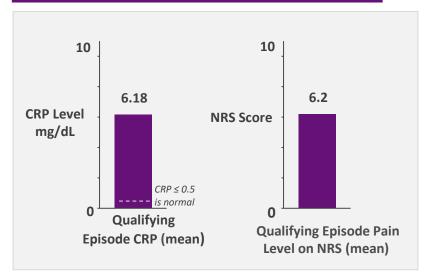
Prior Pericarditis History at Baseline (n=86)



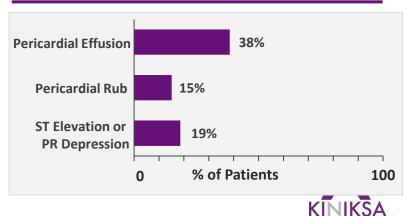




Qualifying Episode CRP & NRS (n=86)

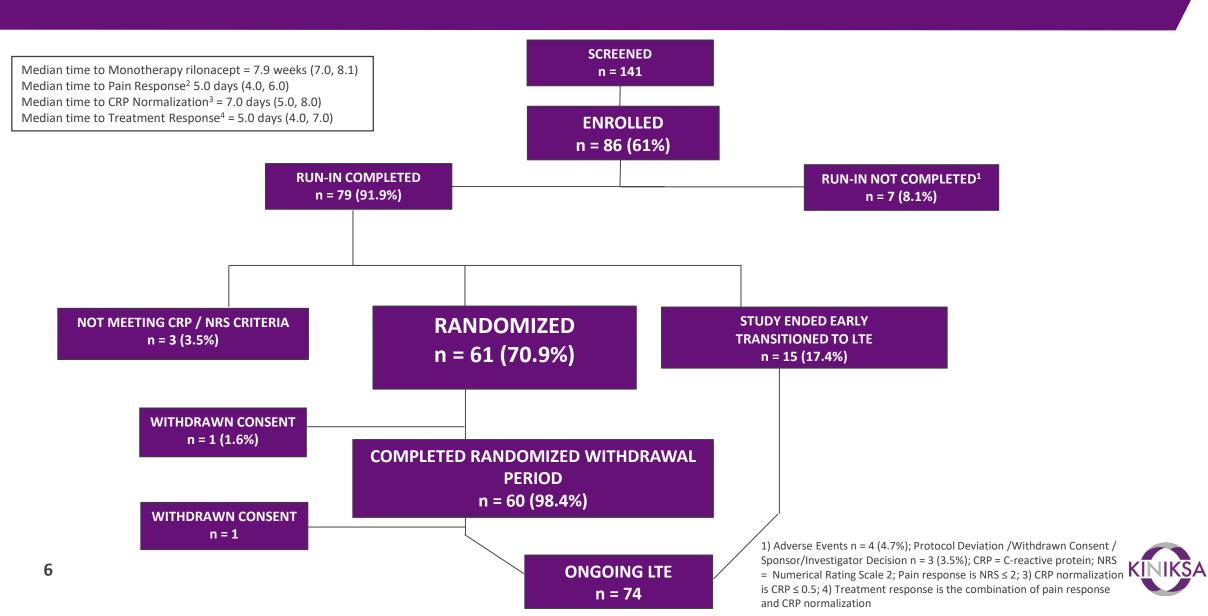


Pericarditis Manifestations at Qualifying Episode (n=86)

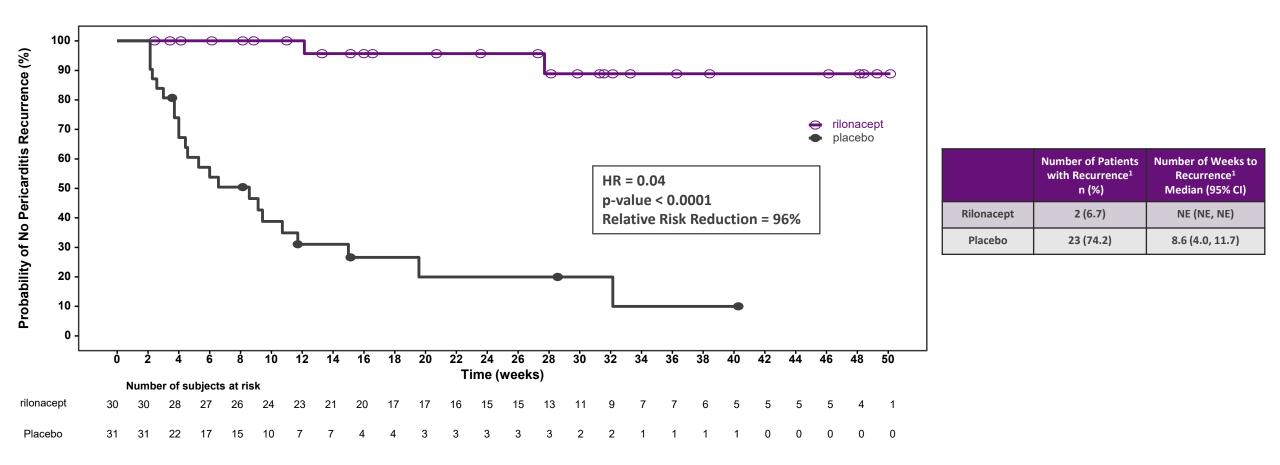


Subject Disposition Pivotal Phase 3 Rilonacept Data





Primary Efficacy Endpoint: Time-to-First Adjudicated Pericarditis Recurrence Pivotal Phase 3 Rilonacept Data



Annualized incidence of pericarditis recurrence decreased from 4.42 episodes per year prior to the study to 0.15 episodes per year while on rilonacept treatment.



RHAPSODY

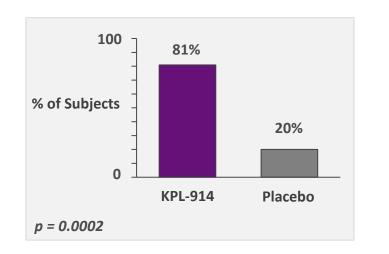
1) Mean (median, range); NE = Not Estimable

7

Secondary Endpoints at Week 16 of the Randomized Withdrawal Period Pivotal Phase 3 Rilonacept Data



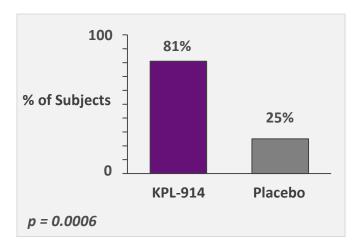
Proportion of Subjects Who Maintained Clinical Response ¹



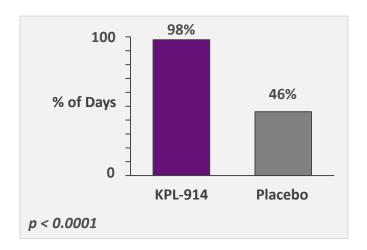
Data at Weeks 8 and 24 were consistent and statistically significant (Week 8, p < 0.0001; Week 24, p=0.0022)

8

Proportion of Subjects with Absent/Minimal Pericarditis Symptoms based on the 7-point PGIPS²



Data at Weeks 8 and 24 were consistent and statistically significant (Week 8, p < 0.0001; Week 24, p=0.0002) Percent of Days with No or Minimal Pain in First 16 Weeks (ITT Week 16)³



Data at Weeks 8 and 24 were consistent and statistically significant (Week 8, p < 0.0001; Week 24, p < 0.0001)

1) Clinical Response is defined as a weekly average of daily pericarditis pain of <2.0 on the 11-point NRS, CRP level <0.5 mg/dL, and on monotherapy of randomized study drug in that week. Subjects who had recurrence, or used bailout rilonacept, or used rescue medication, discontinued double-blinded treatment, or lost to follow-up before the week will be considered as non-responders;

2) PGIPS = Patient Global Impression of Pericarditis Severity baseline;

3) No or minimal pain is defined as non-missing daily NRS ≤ 2. The percentage of days with no or minimal pain in the first 24, 16, and 8 weeks is calculated for each subject using 24x7, 16x7, 8x7, respectively, as the denominator. Missing values in pain diary will be counted as 0 day with no or minimal pain. On days of using ORT or corticosteroid, count as 0 day with no or minimal pain. If bailout rilonacept was used, each administration (loading dose or not) will be counted as 7 days without qualifying no or minimal pain.



Pivotal Phase 3 Rilonacept Data

9



	Run-In Period	Randomized Withdrawal Period		
Category ¹	KPL-914 (N=86) n (%)	KPL-914 Including Bailout Rilonacept (N=30) n (%)	Placebo Only Before Bailout Rilonacept (N=31) n (%)	
All Adverse Events	69 (80.2)	24 (80.0)	13 (41.9)	
TEAEs ²	69 (80.2)	24 (80.0)	13 (41.9)	
TEAEs by Maximum severity ³				
Mild	52 (60.5)	16 (53.3)	9 (29.0)	
Moderate	15 (17.4)	8 (26.7)	4 (12.9)	
Severe	2 (2.3)	0	0	
Drug-Related TEAEs ⁴	46 (53.5)	10 (33.3)	1 (3.2)	
Serious TEAEs (SAE)⁵	1 (1.2)	1 (3.3)	1 (3.2)	
TEAEs Leading to Death	0	0	0	
Drug-Related SAE ⁴	0	0	0	
TEAEs Leading to Dose Interruption	0	1 (3.3)	0	
TEAEs Leading to Study Drug Discontinuation	4 (4.7) ⁶	0	0	
TEAEs of Special Interest (Malignancy) ⁷	0	1 (3.3)	0	
TEAEs of Injection Site Reaction	28 (32.6)	6 (20.0)	0	
TEAEs of Injections and Infestations	14 (16.3)	12 (40.0)	3 (9.7)	

	Run-In Period	Randomized Withdrawal Period		
Category ¹	KPL-914 (N=86) n (%)	KPL-914 Including Bailout Rilonacept (N=30) n (%)		
Bronchitis	0	1 (3.3)	0	
Conjunctivitis	0	1 (3.3)	0	
Ear infection	0	0	0	
Gastroenteritis	0	0	1 (3.2)	
Gastroenteritis viral	0	0	0	
Gastroenteritis viral infection	0	1 (3.3)	1 (3.2)	
Hordeolum	1 (1.2)	0	0	
Influenza	1 (1.2)	0	1 (3.2)	
Nasopharyngitis	6 (7.0)	2 (6.7)	0	
Oral herpes	1 (1.2)	1 (3.3)	0	
Otitis media	0	1 (3.3)	0	
Pharyngitis	1 (1.2)	0	0	
Pharyngitis streptococcal	0	0	0	
Rhinitis	1 (1.2)	0	0	
Sinusitis	1 (1.2)	3 (10.0)	0	
Subcutaneous abscess	1 (1.2)	0	0	
Upper respiratory tract infection	2 (2.3)	1 (3.3)	0	
Urinary tract infection	1 (1.2)	3 (10.0)	0	
Vaginal infection	0	1 (3.3)	0	
Viral upper respiratory tract infection	2 (2.3)	1 (3.3)	0	

1) Subjects with multiple events are counted once in the same category; 2) A Treatment-emergent adverse events (TEAEs) are defined as AEs that start or increase in severity on or after the date of first dose and before 6 weeks after the last dose of study drug; 3) Each subject has only been represented with the maximum severity; 4) Related or possibly related or missing, as assessed by the investigator; 5) SAEs (all unrelated to study drug) - Run in Period: CVA (carotid dissection); RW Period: Chest fluttering after alcohol (on PBO), and Pyrexia, Squamous cell Carcinoma, and post-operative ileus (on rilonacept); 6) alopecia, allergic alveolitis (related to other factors), erythema, and systemic allergic reaction (hypersensitivity); 7) Includes malignancy, excluding basal cell carcinoma of the skin



Recurrent Pericarditis U.S. Prevalence Estimated to be ~40K Patients

~14K patients with inadequate response to conventional therapy and persistent underlying disease

	Clear Call to Action: ~1	4K Patients	
Refractory ^{1,2}	Patient fails to respond, or is intolerant, to NSAIDs, colchicine, and steroids (~3K) or patient fails to respond to NSAIDs and colchicine and not suitable for steroids (~5K)	~8K	Represented by the patient population studied in RHAPSODY
Multiple Relapsing ^{1,2}	Patient previously responding to NSAIDs, colchicine, and/or steroids, but who continues to experience multiple recurrences	~5К	 Highest unmet needs: Resolution of episodes Prevention of future episodes Steroid-sparing disease control Quality of life
Steroid- Dependent ^{1,2}	Patient is unable to be tapered off steroids without experiencing subsequent recurrences	~1K	Physicians indicated an interest to treat across all three subgroups ³

Potential to Broaden Utilization Over Time: ~3K Patients

First Recurrence, High Risk^{1,2} Patient identified during first recurrence as having high risk features predictive of multiple future recurrences (very large effusions, tamponade, etc.)

~3K



1) Klein A, Cremer P, Kontzias A, Furqan M, Tubman R, Roy M, Magestro M. Annals of Epidemiology. 2019;36:71; 2) Lin D, Majeski C, DerSarkissian M, Magestro M, Cavanaugh C, Laliberte F, Lejune D, Mahendran M, Duh M, Klein A, Cremer P, Kontzias A, Furqan M, Tubman R, Roy M, Mage. (Nov, 2019). Real-World Clinical Characteristics and Recurrence Burden of Patients Diagnosed with Recurrent Pericarditis in the United States. Poster session presented at the American Heart Association, Philadelphia, PA.; 3) ClearView Forecasting Analysis 2019 Q1



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Appendix

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Pivotal Phase 3 Rilonacept Data



	Run-In Period	Randomized Withdrawal Pe		
Category ¹	KPL-914 (N=86) n (%)	KPL-914 Including Bailout Rilonacept (N=30) n (%)	Placebo Only Before Bailout Rilonacept (N=31) n (%)	
Subjects with Any TEAEs	69 (80.2)	24 (80.0)	13 (41.9)	
Blood and lymphatic system disorders	2 (2.3)	0	0	
Eosinophilia	1 (1.2)	0	0	
Lymphadenopathy	1 (1.2)	0	0	
Cardiac disorders	5 (5.8)	0	2 (6.5)	
Angina pectoris	1 (1.2)	0	0	
Aortic valve incompetence	0	0	1 (3.2)	
Atrial fibrillation	1 (1.2)	0	0	
Cardiac flutter	0	0	1 (3.2)	
Palpitations	1 (1.2)	0	0	
Sinus tachycardia	1 (1.2)	0	0	
Tachycardia	1 (1.2)	0	0	
Ventricular dyssynchrony	1 (1.2)	0	0	
Ear and labyrinth disorders	1 (1.2)	0	0	
Middle ear effusion	0	0	0	
Vertigo	1 (1.2)	0	0	
Endocrine disorders	0	1 (3.3)	0	
Hypothyroidism	0	1 (3.3)	0	
Eye disorders	1 (1.2)	0	0	
Diplopia	0	0	0	
Eye inflammation	1 (1.2)	0	0	
Gastrointestinal disorders	14 (16.3)	2 (6.7)	2 (6.5)	

	Run-In Period	Randomized V	Vithdrawal Period
Category ¹	KPL-914 (N=86) n (%)	KPL-914 Including Bailout Rilonacept (N=30) n (%)	Placebo Only Before Bailout Rilonacept (N=31) n (%)
Abdominal distension	2 (2.3)	0	0
Abdominal pain	0	0	1 (3.2)
Abdominal tenderness	0	1 (3.3)	0
Aphthous ulcer	0	1 (3.3)	0
Constipation	1 (1.2)	0	0
Diarrhea	5 (5.8)	0	0
Gastric ulcer	1 (1.2)	0	0
Gastritis	1 (1.2)	0	0
Gastrointestinal disorder	1 (1.2)	0	0
Gastrooesophageal reflux disease	1 (1.2)	1 (3.3)	0
Gingival pain	1 (1.2)	0	0
Haemorrhoids	0	0	1 (3.2)
lleus	0	0	0
Nausea	2 (2.3)	0	0
Tongue ulceration	0	1 (3.3)	0
Vomiting	1 (1.2)	0	0
General disorders and administration site conditions	30 (34.9)	10 (33.3)	1 (3.2)
Asthenia	2 (2.3)	0	0
Chest discomfort	1 (1.2)	1 (3.3)	0
Chills	1 (1.2)	0	0
Fatigue	2 (2.3)	2 (6.7)	0
Feeling abnormal	1 (1.2)	0	0



Pivotal Phase 3 Rilonacept Data



	Run-In Period	Randomized Withdrawal Period	
Category ¹	KPL-914 (N=86) n (%)	KPL-914 Including Bailout Rilonacept (N=30) n (%)	Placebo Only Before Bailout Rilonacept (N=31) n (%)
Feeling hot	2 (2.3)	0	0
Injection site bruising	1 (1.2)	0	0
Injection site discolouration	2 (2.3)	0	0
Injection site erythema	18 (20.9)	6 (20.0)	0
Injection site inflammation	1 (1.2)	0	0
Injection site nodule	1 (1.2)	0	0
Injection site pain	4 (4.7)	0	0
Injection site pruritus	5 (5.8)	5 (16.7)	0
Injection site rash	3 (3.5)	0	0
Injection site reaction	2 (2.3)	0	0
Injection site swelling	5 (5.8)	1 (3.3)	0
Non-cardiac chest pain	1 (1.2)	3 (10.0)	1 (3.2)
Oedema peripheral	0	1 (3.3)	0
Pain	1 (1.2)	1 (3.3)	0
Pyrexia	1 (1.2)	0	0
Immune system disorders	1 (1.2)	0	1 (3.2)
Drug hypersensitivity	1 (1.2)	0	0
Hypersensitivity	1 (1.2)	0	0
Seasonal allergy	0	0	1 (3.2)
Infections and infestations	14 (16.3)	12 (40.0)	3 (9.7)
Bronchitis	0	1 (3.3)	0
Conjunctivitis	0	1 (3.3)	0

	Run-In Period	Randomized V	Nithdrawal Period
Category ¹	KPL-914 (N=86) n (%)	KPL-914 Including Bailout Rilonacept (N=30) n (%)	Placebo Only Before Bailout Rilonacept (N=31) n (%)
Ear infection	0	0	0
Gastroenteritis	0	0	1 (3.2)
Gastroenteritis viral	0	0	0
Gastrointestinal viral infection	0	1 (3.3)	1 (3.2)
Hordeolum	1 (1.2)	0	0
Influenza	1 (1.2)	0	1 (3.2)
Nasopharyngitis	6 (7.0)	2 (6.7)	0
Oral herpes	1 (1.2)	1 (3.3)	0
Otitis media	0	1 (3.3)	0
Pharyngitis	1 (1.2)	0	0
Pharyngitis streptococcal	0	0	0
Rhinitis	1 (1.2)	0	0
Sinusitis	1 (1.2)	3 (10.0)	0
Subcutaneous abscess	1 (1.2)	0	0
Upper respiratory tract infection	2 (2.3)	1 (3.3)	0
Urinary tract infection	1 (1.2)	3 (10.0)	0
Vaginal infection	0	1 (3.3)	0
Viral upper respiratory tract infection	2 (2.3)	1 (3.3)	0
Injury, poisoning and procedural complications	6 (7.0)	3 (10.0)	1 (3.2)
Epicondylitis	0	1 (3.3)	0
Fall	2 (2.3)	0	0
Humerus fracture	0	0	1 (3.2)



Pivotal Phase 3 Rilonacept Data



	Run-In Period	Randomized V	Vithdrawal Period		Run-In Period	Randomized V	Vithdrawal Period
Category ¹	KPL-914 (N=86) n (%)	KPL-914 Including Bailout Rilonacept (N=30) n (%)	Placebo Only Before Bailout Rilonacept (N=31) n (%)	Category ¹	KPL-914 (N=86) n (%)	KPL-914 Including Bailout Rilonacept (N=30) n (%)	Placebo Only Before Bailout Rilonacept (N=31) n (%)
Joint injury	0	1 (3.3)	0	Liver function test increased	1 (1.2)	0	0
Limb injury	0	0	1 (3.2)	Low density lipoprotein increased	1 (1.2)	0	0
Muscle strain	1 (1.2)	0	0	Mean cell volume increased	0	1 (3.3)	0
Post procedural contusion	0	1 (3.3)	0	Smear cervix abnormal	1 (1.2)	0	0
Post-traumatic pain	2 (2.3)	0	0	Weight increased	1 (1.2)	0	0
Procedural dizziness	1 (1.2)	0	0	Metabolism and nutrition disorders	0	1 (3.3)	0
Investigations	12 (14.0)	7 (23.3)	0	Hyperlipidaemia	0	1 (3.3)	0
Bacterial test	0	0	0	Musculoskeletal and connective tissue disorders	26 (30.2)	6 (20.0)	4 (12.9)
Blood cholesterol increased	0	1 (3.3)	0	Arthralgia	8 (9.3)	1 (3.3)	0
Blood glucose decreased	0	1 (3.3)	0	Arthritis	0	1 (3.3)	0
Blood glucose increased	1 (1.2)	0	0	Axillary mass	0	1 (3.3)	0
Blood pressure increased	1 (1.2)	1 (3.3)	0	Back pain	3 (3.5)	1 (3.3)	0
Blood triglycerides increased	0	1 (3.3)	0	Groin pain	1 (1.2)	0	0
Body temperature decreased	1 (1.2)	0	0	Joint stiffness	2 (2.3)	0	0
C-reactive protein increased	1 (1.2)	2 (6.7)	0	Musculoskeletal chest pain	3 (3.5)	1 (3.3)	4 (12.9)
Eosinophil count increased	1 (1.2)	0	0	Musculoskeletal pain	3 (3.5)	0	0
Haemoglobin decreased	1 (1.2)	0	0	Myalgia	9 (10.5)	1 (3.3)	0
Heart rate increased	1 (1.2)	1 (3.3)	0	Neck pain	1 (1.2)	0	1 (3.2)
Hepatic enzyme increased	1 (1.2)	1 (3.3)	0	Osteoarthritis	1 (1.2)	0	0
Heart density lipoprotein	1 (1.2)	0	0	Pain in extremity	1 (1.2)	0	0
decreased Heart density lipoprotein				Neoplasms benign, malignant and unspecified (incl cysts and polyps)	1 (1.2)	2 (6.7)	0
increased	0	3 (10.0)	0	Acrochordon	1 (1.2)	0	0
Lipids increased	0	2 (6.7)	0				



15

Pivotal Phase 3 Rilonacept Data



	Run-In Period	Randomized Withdrawal Period	
Category ¹	KPL-914 (N=86) n (%)	KPL-914 Including Bailout Rilonacept (N=30) n (%)	Placebo Only Before Bailout Rilonacept (N=31) n (%)
Lipoma	0	1 (3.3)	0
Squamous cell carcinoma	0	1 (3.3)	0
Nervous system disorders	14 (16.3)	2 (6.7)	0
Carpal tunnel syndrome	1 (1.2)	0	0
Cerebrovascular accident	1 (1.2)	0	0
Dizziness	2 (2.3)	1 (3.3)	0
Dysgeusia	1 (1.2)	0	0
Head discomfort	0	1 (3.3)	0
Headache	7 (8.1)	0	0
Migraine	1 (1.2)	0	0
Presyncope	1 (1.2)	0	0
Somnolence	1 (1.2)	0	0
Psychiatric disorders	1 (1.2)	0	1 (3.2)
Insomnia	0	0	1 (3.2)
Sleep disorder	1 (1.2)	0	0
Renal and urinary disorders	0	1 (3.3)	1 (3.2)
Nephrolithiasis	0	1 (3.3)	0
Renal colic	0	0	1 (3.2)
Reproductive system and breast disorders	1 (1.2)	1 (3.3)	1 (3.2)
Ovarian cyst	1 (1.2)	0	0
Uterine haemorrhage	0	1 (3.3)	0
Uterine polyp	0	0	1 (3.2)

	Run-In Period	Randomized V	Vithdrawal Period
Category ¹	KPL-914 (N=86) n (%)	KPL-914 Including Bailout Rilonacept (N=30) n (%)	Placebo Only Before Bailout Rilonacept (N=31) n (%)
Respiratory, thoracic and mediastinal disorders	15 (17.4)	7 (23.3)	1 (3.2)
Alveolitis allergic	1 (1.2)	0	0
Cough	5 (5.8)	1 (3.3)	0
Dysphonia	0	1 (3.3)	0
Dyspnoea	1 (1.2)	1 (3.3)	0
Epistaxis	1 (1.2)	0	0
Nasal congestion	0	0	0
Oropharyngeal pain	1 (1.2)	3 (10.0)	0
Pharyngeal hypoaesthesia	1 (1.2)	0	0
Respiratory tract congestion	2 (2.3)	0	1 (3.2)
Rhinorrhoea	1 (1.2)	0	0
Sinus congestion	2 (2.3)	2 (6.7)	0
Skin and subcutaneous tissue disorders	11 (12.8)	0	1 (3.2)
Acne	1 (1.2)	0	0
Alopecia	1 (1.2)	0	0
Angioedema	1 (1.2)	0	0
Erythema	2 (2.3)	0	0
Pruritus	2 (2.3)	0	0
Pruritus generalised	2 (2.3)	0	1 (3.2)
Rash	1 (1.2)	0	0
Rash macular	3 (3.5)	0	0
Social circumstances	0	1 (3.3)	0



RHAPSODY

Pivotal	Phase 3	Rilonacept	Data
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	Run-In Period	Randomized Withdrawal Period		
Category ¹	KPL-914 (N=86) n (%)	KPL-914 Including Bailout Rilonacept (N=30) n (%)	Placebo Only Before Bailout Rilonacept (N=31) n (%)	
Menopause	0	1 (3.3)	0	
Vascular disorders	2 (2.3)	1 (3.3)	1 (3.2)	
Hypertension	2 (2.3)	1 (3.3)	1 (3.2)	

	Run-In Period	Randomized Withdrawal Period	
Category ¹	KPL-914 (N=86) n (%)	KPL-914 Including Bailout Rilonacept (N=30) n (%)	Placebo Only Before Bailout Rilonacept (N=31) n (%)
Subjects with Any Serious TEAE	1 (1.2)	1 (3.3)	1 (3.2)
Cardiac disorders	0	0	1 (3.2)
Cardiac flutter	0	0	1 (3.2)
Gastrointestinal disorders	0	0	0
lleus	0	0	0
General disorders and administration site conditions	0	0	0
Pyrexia	0	0	0
Neoplasms benign, malignant and unspecified (incl cysts and polyps)	0	1 (3.3)	0
Squamous cell carcinoma	0	1 (3.3)	0
Nervous system disorders	1 (1.2)	0	0
Cerebrovascular accident	1 (1.2)	0	0

