InflaRx Announces Positive Data from Phase II IXCHANGE Study with Vilobelimab in ANCA-associated Vasculitis (AAV)

- Proof of concept established for potential of vilobelimab to reduce use of corticosteroids in AAV patients: vilobelimab demonstrated comparable efficacy to standard of care
- Use of vilobelimab instead of glucocorticoids led to a substantially lower observed glucocorticoid toxicity
- Vilobelimab demonstrated a good safety and tolerability profile; vilobelimab only treatment arm had lowest number of reported treatment emergent adverse events

Jena, Germany, November 15, 2021 – InflaRx N.V. (Nasdaq: IFRX), a clinical-stage biopharmaceutical company developing anti-inflammatory therapeutics by targeting the complement system, today announced positive data from the European Phase II IXCHANGE study of vilobelimab, a first-in-class anti-C5a antibody, in patients with ANCA-associated vasculitis (AAV). The study achieved its principal objective, demonstrating comparable clinical response of vilobelimab to standard of care, while significantly reducing the need for glucocorticoid (GC) treatment in this life-threatening indication.

“We are pleased to report that vilobelimab monotherapy has shown similar efficacy compared to a standard dose of glucocorticoids in patients with ANCA-associated vasculitis in this trial,” said Dr. Korinna Pilz, Chief Development Officer. “Vilobelimab treatment led to a considerably reduced use of glucocorticoids, resulting in remarkably less treatment emergent adverse events and a reduction in the glucocorticoid toxicity index.”

“The results of the IXCHANGE trial regarding the evidence that vilobelimab has the potential to induce remission of ANCA-associated vasculitis with significant reduction in the dose and associated toxicity of glucocorticoids are exciting,” said Peter A. Merkel, MD, MPH, Professor of Medicine and Epidemiology at the University of Pennsylvania, Director of the Penn Vasculitis Center, and coordinating investigator for the study. “The strong efficacy and safety data in the trial are quite encouraging for the development of this novel agent for the treatment against this organ- and life-threatening disease.”

The randomized, double-blind, placebo-controlled, two-part, Phase II IXCHANGE study enrolled 57 patients (30 in part 1; 27 in part 2) with AAV throughout Europe. Part 1 of the study compared vilobelimab plus a reduced dose of GC (RDGC) therapy to a standard dose of GC (SDGC) therapy, while part 2 compared vilobelimab alone to SDGC. All patients received

standard of care immunosuppressive therapy (rituximab or cyclophosphamide). After loading doses administered at days 1, 4, and 8, patients were given 800 mg of vilobelimab every two weeks for 16 weeks, followed by an eight-week observation period. The principal objective of the trial was to evaluate the efficacy of vilobelimab treatment as a replacement for GC therapy in patients with Granulomatosis with Polyangiitis (GPA) and Microscopic Polyangiitis (MPA). The primary efficacy evaluation was clinical response, primarily defined based on a 50% reduction in Birmingham Vasculitis Activity Score (BVAS) and no worsening in any body system from baseline. Clinical remission was a secondary endpoint and was defined as the number of patients achieving a BVAS of zero at week 16. The study was not statistically powered to demonstrate non-inferiority of vilobelimab alone compared to a standard dose of GC therapy. Other secondary endpoints included the vasculitis damage index (VDI), estimated glomerular filtration rate (eGFR) and the glucocorticoid toxicity index (GTI).

The mean total accumulative dose of GC administered after the screening period until end of study in the different groups was 541.9 mg in the vilobelimab only group, 3751.3 mg in the SDGC group and 1485.8 mg in the vilobelimab + RDGC group. The cumulative mean GC dose received during the 28 days prior to randomization was comparable amongst the three arms with 1894.7 mg (vilobelimab only), 1750.1 mg (SDGC) and 2438.8 mg (vilobelimab + RDGC), respectively.

Clinical response as well as clinical remission were achieved in comparably high rates in all three arms: Clinical response at week 16 in evaluable patients was observed in 16 out of 18 (88.9%) patients in the treatment group receiving vilobelimab alone; in 22 out of 23 (95.7%) patients receiving SDGC; and in 10 out of 13 (76.9%) patients in the vilobelimab + RDGC group.

Clinical remission at week 16 is summarized in the following table:

Dose Groups **No. of evaluable patients**	**vilobelimab (No GC)** **N=18**	**SDGC** **N=23**	**vilobelimab + RDGC** **N=13**	**Pooled vilobelimab** **N=31**
Clinical Remission at Week 16 [N (%)]	14 (77.8%)	20 (87.0%)	10 (76.9%)	24 (77.4%)
No Clinical Remission [N (%)]	4 (22.2%)	3 (13.0%)	3 (23.1%)	7 (22.6%)
Missing at week 16	0	1	2	2

The GTI composite score at week 16 was substantially lowered in the vilobelimab alone group (mean value of 0.8) when compared to the SDGC group (mean value of 44.9) and the vilobelimab + RDGC group (mean value of 26.1).

Assessment of the VDI at week 16 suggested comparable values between groups with the vilobelimab only group showing the lowest value: vilobelimab only group (1.0), SDGC group (1.5) and vilobelimab + RDGC group (1.9).

eGFR, a secondary endpoint of the study, demonstrated no observed medically meaningful changes in all three arms.

The vilobelimab only group had the lowest number of reported treatment emergent adverse events (TEAEs) as well as related TEAEs as summarized in the following table:

	vilobelimab (No GC) N=18			SDGC N=24			vilobelimab + RDGC N=15		
	Pat N	(Pat %)	Events	Pat N	(Pat %)	Events	Pat N	(Pat %)	Events
TEAEs	16	(88.9%)	81	24	(100.0%)	180	15	(100.0%)	89
Serious TEAEs	5	(27.8%)	5	4	(16.7%)	6	3	(20.0%)	3
TEAEs rated as related to vilobelimab	6	(33.3%)	8	18	(75.0%)	40	6	(40.0%)	9
TEAEs rated as related to GC	7	(38.9%)	21	18	(75.0%)	105	11	(73.3%)	24
Serious TEAEs rated as related to vilobelimab	1	(5.6%)	1	1	(4.2%)	1	0	(0.0%)	0
Serious TEAEs rated as related to GC	2	(11.1%)	2	3	(12.5%)	4	0	(0.0%)	0

There was one fatal event of *pneumocystis jiroveci* pneumonia in the vilobelimab only treatment group. This event started on Day 6 after the patient's randomization in the study and lasted for 30 days until the patient's death. The patient had newly diagnosed GPA and was treated with rituximab prior to and during the screening period and received glucocorticoids as part of the standard premedication regimen. The patient received 3 doses of vilobelimab before onset of the event which led to discontinuation of treatment. The event was judged as unlikely related to vilobelimab by the Company. The patient had received a cumulative dose of 6.57 grams of prednisolone equivalent and 4 doses of rituximab (4 x 740 mg) prior to randomization and prior to onset of the event (38 days). Based on an investigator decision, the patient did not receive the guideline and protocol-recommended antibiotic prophylaxis for *pneumocystis jiroveci* pneumonia, which is a known and described serious infection and potential cause of death for patients diagnosed with and treated for AAV. (Sarica et al., Rheumatology 2020;0:1-9). Another serious adverse event of *pneumocystis jiroveci* pneumonia was detected in the standard GC group in a patient who had undergone a comparable AAV induction therapy.

In May 2021, InflaRx reported positive topline data from the US IXPLORE Phase II study of vilobelimab in AAV. The results indicated that vilobelimab, when given in addition to best standard of care, was well tolerated.

The Company plans to discuss the data from both the US and EU studies with regulatory authorities to determine next steps with the program.

About ANCA-Associated Vasculitis (AAV)

AAV is a rare and life-threatening autoimmune disease in which activation of the complement system, and specifically the generation of larger amounts of C5a, is believed to play a key role in the neutrophil-driven vessel inflammation that defines the disease. AAV affects approximately 40,000 and 75,000 patients in the United States and Europe, respectively.

About vilobelimab:

Vilobelimab is a first-in-class monoclonal anti-human complement factor C5a antibody, which highly and effectively blocks the biological activity of C5a and demonstrates high selectivity towards its target in human blood. Thus, vilobelimab leaves the formation of the membrane attack complex (C5b-9) intact as an important defense mechanism, which is not the case for molecules blocking the cleavage of C5. Vilobelimab has been demonstrated to control the inflammatory response driven tissue and organ damage by specifically blocking C5a as a key “amplifier” of this response in pre-clinical studies. Vilobelimab is believed to be the first monoclonal anti-C5a antibody introduced into clinical development. Over 300 people have been treated with vilobelimab in completed clinical trials, and the antibody has been shown to be well tolerated. Vilobelimab is currently being developed for various inflammatory indications, including hidradenitis suppurativa, ANCA-associated vasculitis and pyoderma gangraenosum, as well as severe COVID-19 and cutaneous squamous cell carcinoma (cSCC).

About InflaRx N.V.:

InflaRx (Nasdaq: IFRX) is a clinical-stage biopharmaceutical company focused on applying its proprietary anti-C5a technology to discover and develop first-in-class, potent and specific inhibitors of C5a. Complement C5a is a powerful inflammatory mediator involved in the progression of a wide variety of autoimmune and other inflammatory diseases. InflaRx was founded in 2007, and the group has offices and subsidiaries in Jena and Munich, Germany, as well as Ann Arbor, MI, USA. For further information please visit www.inflarx.com.

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FORWARD-LOOKING STATEMENTS

This press release contains forward-looking statements. All statements other than statements of historical fact are forward-looking statements, which are often indicated by terms such as “may,” “will,” “should,” “expect,” “plan,” “anticipate,” “could,” “intend,” “target,” “project,” “believe,” “estimate,” “predict,” “potential” or “continue” and similar expressions. Forward-looking statements appear in a number of places throughout this release and may include statements regarding our intentions, beliefs, projections, outlook, analyses and current expectations concerning, among other things, our ongoing and planned clinical trials of vilobelimab as treatment of ANCA-associated vasculitis and the safety and efficacy results of those trials; the impact of the COVID-19 pandemic on the Company; the timing and our ability to commence and conduct clinical trials; potential results from current or potential future collaborations; our ability to make regulatory filings, obtain positive guidance from regulators, and obtain and maintain regulatory approvals for our product candidates; our intellectual property position; our ability to develop commercial functions; expectations regarding clinical trial data; our results of operations, cash needs, financial condition, liquidity, prospects, future transactions, growth and strategies; the industry in which we operate; the trends that may affect the industry or us and the risks, uncertainties and other factors described under the heading “Risk Factors” in InflaRx’s periodic filings with the Securities and Exchange Commission. These statements speak only as of the date of this press release and involve known and unknown risks, uncertainties and other important factors that may cause our actual results, performance or achievements to be materially different from any future results, performance or achievements expressed or implied by the forward-looking statements. Given these risks, uncertainties and other factors, you should not place undue reliance on these forward-looking statements, and we assume no obligation to update these forward-looking statements, even if new information becomes available in the future, except as required by law.