Akari Therapeutics Demonstrates Positive Response with Coversin in Ongoing Phase 2 PNH Trial and In Additional Clinical Targets

-Interim Phase 2 PNH data demonstrate positive response with Coversin™
-Phase 3 PNH program expected to commence in 4Q2017
-Data from preclinical aHUS model demonstrates positive results
-New preclinical data demonstrates positive response of Coversin's combined C5 and LTB4 therapy in skin and eye models
-Phase 2 programs in Mucous Membrane Pemphigoid (eye) and Bullous Pemphigoid (Skin) expected to commence in 1Q2018

-Company to webcast today's Research and Development Day at 8:00 am Eastern (details below)-

NEW YORK and LONDON, April 24, 2017 (GLOBE NEWSWIRE) -- Akari Therapeutics (NASDAQ: AKTX), an emerging growth, clinical-stage biopharmaceutical company, announced that it will present data from an interim analysis of its ongoing Phase 2 trial of Coversin in paroxysmal nocturnal hemoglobinuria (PNH), as well as preclinical data for additional indications and other opportunities, at today's Research and Development Day.

Positive Interim Phase 2 data in PNH

In this 90 day, open label Phase 2 trial conducted at five centers in the EU, five patients with PNH who had not received prior anti-complement therapy were enrolled and treated with Coversin self-administered subcutaneous injections twice a day for approximately the first month and then switched to once daily injections. The primary endpoint in this trial is reduction in serum LDH to ≤1.8 X ULN or 500 I U/L whichever is the lower from day 1 (pre-dose) to day 28. Secondary endpoints are LDH at days 60 and 90, hemoglobin, CH50, quality of life, and transfusion independence. The objectives of our Phase 2 study are to validate the safety and efficacy of Coversin, confirm convenience of our dosing regimen, and study dose ranging to identify the correct treatment dose in advance of Phase 3.

The 4 patients who remain on Coversin are characterized, to date, by:

- Symptom free
- LDH reductions 1.3, 1.4, 1.5 and 1.8X ULN
- No transfusions (2 of the 4 patients received transfusions in the 3 months prior to the study)
- CH50 below level of quantification (from day 1)
- Once daily subcutaneous self-administration
- No neutralizing antibodies
- No serious adverse events (SAEs)

In this dose ranging Phase 2 study, the protocol allowed for patients to be updosed from the 30mg starting dose. Of the 4 patients continuing on Coversin: the first patient's LDH went from 2.4X ULN at baseline to 2.1X ULN on the starting dose, was updosed to 45 mg and achieved a reduction to 1.3X ULN on day 28 and remains on 45mg once daily injections; the second patient with an LDH of 7.5X ULN at baseline, achieved a reduction to 1.4X ULN on day 28 with the starting dose, and remains on 30mg once daily injections; the third patient's LDH went from 3.3X ULN at baseline to 2.4X ULN on the starting dose, was updosed to 45 mg and achieved a reduction to 1.5X ULN on day 60 and remains on 45mg once daily

injections; and the fourth patient who just reached the 6 week mark for this interim analysis achieved an LDH reduction from 5.6 X ULN at baseline to 1.8X ULN on day 40 on the starting dose, and was updosed to 45mg on day 48 and continues on once daily injections. All 4 patients achieved on day 1 and throughout the trial a CH50 below the lower limit of quantification ("<LLQ").

A fifth patient with an LDH of 3.7 X ULN at baseline achieved the primary endpoint at day 14, but was withdrawn from the trial at day 43 due to a suspected co-morbidity unrelated to treatment, which would have excluded the patient from the trial protocol. While on Coversin, the patient met the primary endpoint (day 14), and achieved and maintained a CH50 <LLQ (day 1) but clinical response fluctuated and did not stabilize. After withdrawal, the patient switched to eculizumab. On eculizumab, LDH decreased to below 1.5X ULN and the patient experienced other clinical complications.

As reported previously, an eculizumab-resistant PNH patient had been under treatment with subcutaneous Coversin for over 14 months under an approved clinical protocol. The patient continues to self-administer Coversin and continues to demonstrate complete complement inhibition without any change in dose. The patient's most recent reported LDH was below 1.3 X ULN. Further, there have been no signs of neutralizing antibodies.

All patients are comfortable with self-dosing and by the end of May, we plan to have the four continuing patients from this Phase 2 and the one patient from the eculizumab resistant protocol on long term treatment in our long term open label safety trial. Akari is planning to initiate its Phase 3 program in PNH in the fourth quarter of 2017 and anticipates initial Phase 3 data 1Q2019.

aHUS

Recent studies with Coversin have demonstrated positive results in a preclinical model of atypical hemolytic uremic syndrome (aHUS) conducted by Prof. Giuseppe Remuzzi and colleagues Marina Noris and Miriam Galbusera at the Mario Negri Institute for Pharmacological Research in Bergamo, Italy, and the Clinical Research Center for Rare Diseases "Aldo e Cele Dacco" of the same institute, a European center for the study of aHUS. In a well-established ex vivo model testing sera of patients with aHUS, Coversin demonstrated a statistically significant ($p<0.001$) reduction in membrane attack complex (MAC) deposition on endothelial cells when activated by sera of patients with active aHUS, at least as well as eculizumab. Akari expects to initiate its Phase 2 trial in aHUS in 2Q2017, and anticipates Phase 2 aHUS data 2Q2018.

New data demonstrating Coversin C5 and LTB4 dual activity in eye and skin models

Akari will present new data on its clinical development plan for Coversin based on its dual C5/LTB4 inhibition, focusing on new clinical indications in the eye and skin.

Results in a rodent model of Experimental Immune Conjunctivitis (EIC), undertaken at the world leading Moorfields Hospital Institute of Ophthalmology, showed that Coversin demonstrated significant anti-inflammatory activity with both C5 and LTB4 inhibition believed to play a role. In this preclinical model of severe eye surface inflammation, Coversin, applied topically, resulted in a statistically significant reduction (64%, $p<0.001$) in late phase inflammation versus placebo.

In a pre-clinical mouse model of bullous pemphigoid (a blistering disease of the skin), where both LTB4 and C5 are thought to be dysregulated, Coversin demonstrated a statistically significant reduction (~60%, $p=0.002$) in affected area with Coversin compared to placebo and steroids

Based on these results, Akari, while continuing to develop Coversin in PNH and aHUS, also intends to focus on new indications for Coversin in diseases where both C5 and LTB4 are believed to be involved. Akari expects to commence in 1Q2018 randomized, double blind Phase 2 trials in patients with bullous pemphigoid who are refractory to oral steroids as well in mucous membrane pemphigoid (eye) and anticipates Phase 2 data from these trials in 4Q2018.

Pipeline

Akari will present an update on its pipeline of new molecules including:

- new data in an ex vivo model testing activity in the alternative pathway of Complement of its long acting version of Coversin, PAS Coversin, which is designed for weekly dosing, demonstrated that PAS Coversin could potentially be more potent than Coversin and eculizumab . This data will be presented by Professor Arne Skerra, Professor of Biological Chemistry at the University of Munich and Chairman of XL-protein GmbH. Akari anticipates a Phase 1 trial for PAS Coversin to commence in 3Q2018.
- positive data in a range of ex vivo lung models using a new Akari molecule that binds only to LTB4. This opens up a range of new inflammatory target conditions for Akari where LTB4 dysregulation is potentially the primary pathophysiology of disease. This data will be presented by Dr Robert Snelgrove, National Heart and Lung Institute, Imperial College London whose particular expertise lies in LTB4 and lung inflammation.

Akari anticipates further preclinical pipeline data in 3Q2017.

Webcast

The R&D Day presentation, scheduled to begin at 8:00am EDT today, April 24, 2017, will be webcast live and can be accessed by following the link on the homepage of our website (www.akaritx.com) as well as through the "Link to Simulcast for April 24th R&D Day" which appears on the left side of the "Investor Relations" section of Akari's website.

About Akari Therapeutics Plc

Akari is a clinical-stage biopharmaceutical company focused on the development and commercialization of life-transforming treatments for a range of rare and orphan autoimmune and inflammatory diseases caused by dysregulation of complement C5 and Leukotriene B4 (LTB4), including paroxysmal nocturnal hemoglobinuria ("PNH"), atypical Hemolytic Uremic Syndrome ("aHUS"), and Guillain Barré syndrome ("GBS"). Akari's lead product candidate, Coversin™ complement inhibitor, a second-generation complement inhibitor, acts on complement component-C5, preventing the release of C5a and the formation of C5b–9 (also known as the membrane attack complex or MAC), and independently also

inhibits LTB4 activity. C5 inhibition is growing in importance in a range of rare autoimmune diseases related to dysregulation of the complement component of the immune system, including PNH, aHUS, and GBS. Exploiting the power of nature, Akari is also developing other tick derived proteins and expects to bring additional compounds to clinical trials over the next several years. The pipeline is focused on developing bioengineered versions of native tick salivary proteins that act as anti-inflammatory compounds allowing the tick to remain on its host. These compounds include PGP sparing LTB4 inhibitors, classical and alternative complement inhibitors, anti-histamines, and serotonin inhibitors as examples. Akari is also developing engineered forms that allow for potential oral absorption, as, for example, a potential orally absorbed C5 inhibitor, and tissue specific proteins, as, for example, Coversin™ that acts specifically at the neuromuscular junction for diseases like myasthenia gravis

Cautionary Note Regarding Forward-Looking Statements

Certain statements in this press release constitute "forward-looking statements" within the meaning of the Private Securities Litigation Reform Act of 1995. These forward-looking statements reflect our current views about our plans, intentions, expectations, strategies and prospects, which are based on the information currently available to us and on assumptions we have made. Although we believe that our plans, intentions, expectations, strategies and prospects as reflected in or suggested by those forward-looking statements are reasonable, we can give no assurance that the plans, intentions, expectations or strategies will be attained or achieved. Furthermore, actual results may differ materially from those described in the forward-looking statements and will be affected by a variety of risks and factors that are beyond our control. Such risks and uncertainties for our company include, but are not limited to: needs for additional capital to fund our operations, an inability or delay in obtaining required regulatory approvals for Coversin and any other product candidates, which may result in unexpected cost expenditures; risks inherent in drug development in general; uncertainties in obtaining successful clinical results for Coversin and any other product candidates and unexpected costs that may result therefrom; failure to realize any value of Coversin and any other product candidates developed and being developed in light of inherent risks and difficulties involved in successfully bringing product candidates to market; inability to develop new product candidates and support existing product candidates; the approval by the FDA and EMA and any other similar foreign regulatory authorities of other competing or superior products brought to market; risks resulting from unforeseen side effects; risk that the market for Coversin may not be as large as expected; inability to obtain, maintain and enforce patents and other intellectual property rights or the unexpected costs associated with such enforcement or litigation; inability to obtain and maintain commercial manufacturing arrangements with third party manufacturers or establish commercial scale manufacturing capabilities; the inability to timely source adequate supply of our active pharmaceutical ingredients from third party manufacturers on whom the company depends; our inability to obtain additional capital on acceptable terms, or at all; unexpected cost increases and pricing pressures; uncertainties of cash flows and inability to meet working capital needs; and risks and other risk factors detailed in our public filings with the U.S. Securities and Exchange Commission, including our Annual Report on Form 20-F filed on March 31, 2017. Except as otherwise noted, these forward-looking statements speak only as of the date of this press release and we undertake no obligation to update or revise any of these statements to reflect events or circumstances occurring after this press release. We caution investors not to place considerable reliance on the forward-looking statements contained in this press release.

Contact:
Investor Contact:
The Trout Group
Lee Stern

lstern@troutgroup.com
646–378–2922
Media Contact:
Susan Forman / Laura Radocaj
Dian Griesel Int'l.
(212) 825-3210