Nicox Receives Positive Opinion for European Orphan Drug Designation for Naproxcinod for the Treatment of Duchenne Muscular Dystrophy

September 11, 2013.

Sophia Antipolis, France.

Nicox S.A. (NYSE Euronext Paris: COX) today announced that it has received a positive opinion from the European Union Committee for Orphan Medicinal Products (COMP) recommending orphan drug designation for naproxcinod, a CINOD (Cyclooxygenase-Inhibiting Nitric Oxide-Donating) anti-inflammatory candidate, for the treatment of Duchenne Muscular Dystrophy (DMD). The orphan drug designation allows companies to benefit from a number of incentives, including a 10-year market exclusivity post-approval, scientific advice and fee reductions. Final approval of the opinion on orphan designation is expected from the European Commission in the coming months. Naproxcinod has shown promising preclinical results in models of muscular dystrophy.

"*Naproxcinod may have the potential to address the important unmet needs of the patients suffering from Duchenne Muscular Dystrophy, who currently have no approved therapies available."* said **Michele Garufi, Chairman and CEO of Nicox**. *"Receiving the orphan drug designation for naproxcinod in Europe will be a key milestone for Nicox and will support our strategy of partnering the development of naproxcinod in this debilitating disease*."

Nicox is evaluating the options for developing naproxcinod through a partner as an adjuvant for the treatment of muscular dystrophy or for the treatment of the signs and symptoms of osteoarthritis of the knee. This approach is aimed at maximising the opportunities to progress the development of naproxcinod in one of these indications.

About naproxcinod in DMD

In DMD, muscle damage is caused by the inability to synthesize the dystrophin protein, which is associated with neuronal NO-synthase. Naproxcinod, through both its anti-inflammatory and NO-donating properties, may have the potential to bring valuable therapeutic activity to the dystrophic muscles of Duchenne patients.

In a long-term preclinical study, naproxcinod was shown to improve skeletal and cardiac muscle function and to reduce skeletal muscle inflammation in *mdx* mice. The data was presented at the Muscular Dystrophy Association (MDA) Scientific Conference in Washington, DC in April 2013.

A patent covering nitric oxide releasing compounds, including naproxcinod, for the treatment of muscular dystrophies is pending in Europe and has been granted in the United States in July 2013.

About Duchenne Muscular Dystrophy (DMD)

DMD is a chronically debilitating and life-threatening disease, characterized by rapidly progressive muscle weakness and wasting due to degeneration of skeletal, smooth and cardiac muscles. DMD is the most common and serious form of muscular dystrophy, with the onset of symptoms occurring in early childhood (usually between three and five years of age), and affects about 3 in 10,000 live-born males. This condition worsens throughout childhood, with patients becoming wheelchair-bound between the ages of seven and thirteen. Most DMD patients die by the age of twenty, most frequently as a direct result of respiratory/cardiac failure.

About Orphan Drug Designation

The Orphan Medicinal Product Designation is given to products to be developed for the diagnosis, prevention or treatment of life-threatening or very serious conditions that affect not more than 5 in 10,000 persons in the European Union.

This press release contains certain forward-looking statements. Although the Company believes its expectations are based on reasonable assumptions, these forward-looking statements are subject to numerous risks and uncertainties, which could cause actual results to differ materially from those anticipated in the forward-looking statements.

Risks factors which are likely to have a material effect on Nicox's business are presented in the 4th chapter of the « Document de référence, rapport financier annuel et rapport de gestion 2012 » filed with the French Autorité

des Marchés Financiers (AMF) on March 22, 2013 and available on Nicox's website (www.nicox.com) and on the AMF's website (www.amf-france.org).

Nicox Contacts

Nicox	**Gavin Spencer** | **Executive Vice President Corporate Development** Tel +33 (0)4 97 24 53 00 | communications@nicox.com
Media Relations Europe	**FTI Consulting** Julia Phillips | D+44 (0)20 7269 7187 | M +44 (0) 7770 827 263 Julia.Phillips@fticonsulting.com Stephanie Cuthbert | D +44 (0)20 3077 0458 | M +44 (0) 7843 080947 Stephanie.Cuthbert@fticonsulting.com Mo Noonan | D +44 (0)20 7269 7116 | M +44 (0)7876 444 977 Mo.Noonan@fticonsulting.com