

Lexicon and Ipsen enter into ex-North America/Japan licensing and commercialization agreement for telotristat etiprate

- **Telotristat etiprate, a Phase 3 compound for the treatment of carcinoid syndrome**
- **Lexicon to potentially receive \$145 million in upfront and milestone payments, plus royalties**

Paris (France) and the Woodlands, Texas (United States), 22 October 2014 – Ipsen (Euronext: IPN; ADR: IPSEY) and Lexicon Pharmaceuticals, Inc. (Nasdaq: LXX) today announced that they have entered into an exclusive licensing agreement for Ipsen to commercialize telotristat etiprate outside of North America and Japan, with a focus on the treatment of carcinoid syndrome. Lexicon retains sole rights to commercialize telotristat etiprate in the United States, Canada and Japan.

Lexicon is conducting Phase 3 clinical trials of telotristat etiprate for carcinoid syndrome, a serious condition caused by symptomatic neuroendocrine tumors, which produce large amounts of serotonin. Carcinoid syndrome is characterized by severe diarrhea, flushing and, in some cases, heart valve damage. Telotristat etiprate is an oral, small-molecule inhibitor of tryptophan hydroxylase (TPH) that reduces peripheral serotonin production without affecting brain serotonin levels. Telotristat etiprate has received fast track status and orphan drug designation from the Food and Drug Administration in the United States, and has received orphan drug designation from the European Medicines Agency.

Lexicon will continue to lead the global Phase 3 clinical program for telotristat etiprate in carcinoid syndrome, from which data are expected in 2015. The pivotal Phase 3 trial is comparing telotristat etiprate to placebo on a background of somatostatin analog (SSA) therapy, the current standard of care, in patients whose carcinoid syndrome is not adequately controlled with lanreotide or octreotide. The clinical Phase 3 study is recruiting in approximately 70 centers worldwide. Lexicon will continue to be responsible for the potential registration of telotristat etiprate in the U.S., Canada and Japan, while Lexicon and Ipsen will collaborate to seek regulatory approvals in Europe and other countries within the Ipsen licensed territory, with Ipsen assuming the lead responsibility in those markets.

“This collaboration with Ipsen provides the opportunity to create added value for Lexicon and more effectively commercialize telotristat etiprate in markets around the world.” said **Lyonel**



Coats, Lexicon's President and Chief Executive Officer. *"Ipsen is a leader in the treatment of carcinoid syndrome and neuroendocrine tumors with Somatuline[®] (lanreotide). Lexicon will benefit commercially from Ipsen's substantial market presence in Europe and other countries in the licensed territory, and will benefit globally from coordination with Ipsen on medical and scientific matters. This agreement represents an important achievement for Lexicon as we continue to execute on a strategy to translate our discoveries into life-changing products for patients in important areas of unmet medical need."*

Marc de Garidel, Chairman and Chief Executive Officer of Ipsen stated: *"Ipsen is delighted to enter into a partnership with Lexicon in Europe and other countries. The CLARINET[®] study with Somatuline[®] (lanreotide) paved the way for a change in the treatment paradigm for pancreatic and gastro intestinal NET patients. Upon approval, telotristat etiprate will enlarge our footprint in the symptomatic management of NET and offer important benefits to patients with carcinoid syndrome."* **Marc de Garidel** added: *"This agreement reflects our strategy to strengthen our presence in the fields of oncology, endocrinology and neurology, which will continue to be supported by our newly signed five-year multi-currency €500 million revolving credit facility."*

Under the financial terms of the agreement, Lexicon is eligible to receive up to \$145 million, comprising \$23 million upfront payment and additional payments contingent upon achievement of clinical, regulatory and commercial milestones. In addition, Lexicon is also eligible to receive royalties on net sales of telotristat etiprate in the licensed territory.

About carcinoid syndrome

Neuroendocrine neoplasms are tumors arising from the diffuse neuroendocrine system most often along the gastrointestinal tract. They are rare but their incidence is increasing (approximately 2.5 to 4.5 new cases diagnosed per 100,000 persons per year). They constitute a heterogeneous group of tumors with location of the primary tumor in the gastric mucosa, pancreas, small and large intestine. As a result of their origin, they are capable of releasing a variety of hormones and neuroamines, most commonly serotonin, which, when released into the systemic circulation, can cause distinct clinical symptoms, such as the carcinoid syndrome associating diarrhea and flushing, with often abdominal pain. In some cases, digestive disorders and flushing are associated with heart failure (wheezing, edema of the lower limbs, irregular heartbeats) indicator of carcinoid valvular heart disease.

Lexicon Conference Call

Lexicon management will hold a conference call at 8:30 a.m. Eastern Time on October 22, 2014. The dial-in number for the conference call is 888-645-5785 (within the US/Canada) or 970-300-1531 (international). The conference ID for all callers is 11632. Investors can access a live webcast of the call at www.lexpharma.com. An archived version of the webcast will be available on the website through November 22, 2014.



About Lexicon

Lexicon is a biopharmaceutical company focused on developing breakthrough treatments for human disease. Lexicon has clinical-stage drug programs for diabetes, carcinoid syndrome, and other indications, all of which were discovered by Lexicon's research team. Lexicon has used its proprietary gene knockout technology to identify more than 100 promising drug targets. For additional information about Lexicon and its programs, please visit www.lexpharma.com.

About Ipsen

Ipsen is a global specialty-driven pharmaceutical company with total sales exceeding €1.2 billion in 2013. Ipsen's ambition is to become a leader in specialty healthcare solutions for targeted debilitating diseases. Its development strategy is supported by 3 franchises: neurology, endocrinology and uro-oncology. Moreover, the Group has an active policy of partnerships. Ipsen's R&D is focused on its innovative and differentiated technological platforms, peptides and toxins. In 2013, R&D expenditure totaled close to €260 million, representing more than 21% of Group sales. Moreover, Ipsen also has a significant presence in primary care. The Group has close to 4,600 employees worldwide. Ipsen's shares are traded on segment A of Euronext Paris (stock code: IPN, ISIN code: FR0010259150) and eligible to the "Service de Règlement Différé" ("SRD"). The Group is part of the SBF 120 index. Ipsen has implemented a Sponsored Level I American Depositary Receipt (ADR) program, which trade on the over-the-counter market in the United States under the symbol IPSEY. For more information on Ipsen, visit www.ipсен.com.

Lexicon Forward Looking Statements

This press release contains "forward-looking statements," including statements relating to Lexicon's clinical development of telotristat etiprate, characterizations of the results of and projected timing of clinical trials of telotristat etiprate, and the potential therapeutic and commercial potential of telotristat etiprate. The press release also contains forward-looking statements relating to Lexicon's growth and future operating results, discovery and development of products, strategic alliances and intellectual property, as well as other matters that are not historical facts or information. All forward-looking statements are based on management's current assumptions and expectations and involve risks, uncertainties and other important factors, specifically including those relating to Lexicon's ability to meet its capital requirements, successfully conduct clinical development of telotristat etiprate and preclinical and clinical development of its other potential drug candidates, advance additional candidates into preclinical and clinical development, obtain necessary regulatory approvals, achieve its operational objectives, obtain patent protection for its discoveries and establish strategic alliances, as well as additional factors relating to manufacturing, intellectual property rights, and the therapeutic or commercial value of its drug candidates, that may cause Lexicon's actual results to be materially different from any future results expressed or implied by such forward-looking statements. Information identifying such important factors is contained under "Risk Factors" in Lexicon's annual report on Form 10-K for the year ended December 31, 2013, as filed with the Securities and Exchange Commission. Lexicon undertakes no obligation to update or revise any such forward-looking statements, whether as a result of new information, future events or otherwise.

Ipsen Forward Looking Statements

The forward-looking statements, objectives and targets contained herein are based on the Group's management strategy, current views and assumptions. Such statements involve known and unknown

risks and uncertainties that may cause actual results, performance or events to differ materially from those anticipated herein. All of the above risks could affect the Group's future ability to achieve its financial targets, which were set assuming reasonable macroeconomic conditions based on the information available today. Use of the words "believes," "anticipates" and "expects" and similar expressions are intended to identify forward-looking statements, including the Group's expectations regarding future events, including regulatory filings and determinations. Moreover, the targets described in this document were prepared without taking into account external growth assumptions and potential future acquisitions, which may alter these parameters. These objectives are based on data and assumptions regarded as reasonable by the Group. These targets depend on conditions or facts likely to happen in the future, and not exclusively on historical data. Actual results may depart significantly from these targets given the occurrence of certain risks and uncertainties, notably the fact that a promising product in early development phase or clinical trial may end up never being launched on the market or reaching its commercial targets, notably for regulatory or competition reasons. The Group must face or might face competition from generic products that might translate into a loss of market share. Furthermore, the Research and Development process involves several stages each of which involves the substantial risk that the Group may fail to achieve its objectives and be forced to abandon its efforts with regards to a product in which it has invested significant sums. Therefore, the Group cannot be certain that favourable results obtained during pre-clinical trials will be confirmed subsequently during clinical trials, or that the results of clinical trials will be sufficient to demonstrate the safe and effective nature of the product concerned. There can be no guarantees a product will receive the necessary regulatory approvals or that the product will prove to be commercially successful. If underlying assumptions prove inaccurate or risks or uncertainties materialize, actual results may differ materially from those set forth in the forward-looking statements. Other risks and uncertainties include but are not limited to, general industry conditions and competition; general economic factors, including interest rate and currency exchange rate fluctuations; the impact of pharmaceutical industry regulation and health care legislation; global trends toward health care cost containment; technological advances, new products and patents attained by competitors; challenges inherent in new product development, including obtaining regulatory approval; the Group's ability to accurately predict future market conditions; manufacturing difficulties or delays; financial instability of international economies and sovereign risk; dependence on the effectiveness of the Group's patents and other protections for innovative products; and the exposure to litigation, including patent litigation, and/or regulatory actions. The Group also depends on third parties to develop and market some of its products which could potentially generate substantial royalties; these partners could behave in such ways which could cause damage to the Group's activities and financial results. The Group cannot be certain that its partners will fulfil their obligations. It might be unable to obtain any benefit from those agreements. A default by any of the Group's partners could generate lower revenues than expected. Such situations could have a negative impact on the Group's business, financial position or performance. The Group expressly disclaims any obligation or undertaking to update or revise any forward looking statements, targets or estimates contained in this press release to reflect any change in events, conditions, assumptions or circumstances on which any such statements are based, unless so required by applicable law. The Group's business is subject to the risk factors outlined in its registration documents filed with the French Autorité des Marchés Financiers.



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