JAZZ PHARMACEUTICALS AND GENTIUM ANNOUNCE EUROPEAN COMMERCIAL LAUNCH OF FIRST APPROVED LIFE-SAVING TREATMENT FOR SEVERE HEPATIC VENO-OCCLUSIVE DISEASE

Defitelio®▼ (defibrotide) Launch Announced During the 40th Annual Meeting of the European Society for Blood and Marrow Transplantation (EBMT) 2014

DUBLIN, Ireland, and VILLA GUARDIA, Italy, 31 March 2014 -- Jazz Pharmaceuticals plc (Nasdaq: JAZZ) and Gentium S.p.A., a Jazz Pharmaceuticals company, today announced the commencement of the European commercial launch of Defitelio®▼ (defibrotide), the first licensed product for the treatment of severe hepatic veno-occlusive disease (severe VOD or sVOD) in patients over one month of age undergoing haematopoietic stem cell transplantation (HSCT) therapy.[1] The companies have launched Defitelio in Germany and Austria and expect to continue the launch in 27 additional European countries on a rolling basis during 2014 and 2015.

Severe VOD, one of the most serious early complications in HSCT therapy, is associated with multi-organ failure and is fatal in over 80% of patients.[2,3] HSCTs are performed with curative intent in patients with haematological malignancies, selected solid tumours and some non-malignant disorders, such as serious haemoglobinopathies.[4,5]

“The commercial availability of Defitelio as the first medicine licensed for the treatment of sVOD in Europe is an important step forward for patients with this life-threatening condition,” said Bruce C. Cozadd, chairman and CEO at Jazz Pharmaceuticals plc. “Additionally, this European launch represents a key milestone for the combined Jazz Pharmaceuticals and Gentium team following the acquisition of Gentium by Jazz Pharmaceuticals earlier this year, and reinforces our commitment to bringing important therapies to patients who have significant unmet medical needs in the areas of haematology and oncology.”

“Severe VOD is a complex and unpredictable disease, and its impact on patients, physicians and resources is substantial. Early and effective intervention is crucial in saving lives and limiting the potentially significant burden of this disease, and physicians have been eagerly awaiting the commercial availability of Defitelio in Europe,” said Professor Mohamad Mohty, President-Elect of the EBMT and Professor of Haematology, Saint-Antoine Hospital and University Pierre & Marie Curie, Paris.

The efficacy of Defitelio to treat sVOD in HSCT patients is supported by data from a pivotal, multi-centre Phase 3 trial that evaluated Defitelio for the treatment of sVOD compared with a historical control group of patients who had received standard supportive care.[1] In this trial, Defitelio was shown to provide a significant increase in survival rates for patients with sVOD in HSCT. The results demonstrated a 52% increase in survival at 100 days after transplantation for patients treated with Defitelio compared to patients in the historical control group (38.2% in the Defitelio group vs. 25.0% in the historical control group; p=0.0341).[1] In the clinical trial, 23.5% of patients treated with Defitelio achieved complete response at 100 days after transplantation versus 9.4% of patients in the historical control group (p=0.013).[1]

The efficacy data from this pivotal trial are supported with data from a Phase 2 dose-finding study, as well as data from the International Compassionate Use Programme and an interim analysis (subset of patients with sVOD) of an ongoing, open-label treatment investigational new drug (IND) study being conducted in the United States (U.S.).[1] Additionally, data derived from

an independent registry in the U.S. supported the European approval of Defitelio for use in patients with sVOD.[1]

Treatment with Defitelio has generally been well tolerated in all age groups.[1,6] In the Phase 3 pivotal trial, the overall incidence of adverse events was similar in the Defitelio treatment group and in the control group.[7] The most frequent adverse events observed during pre-marketing use were haemorrhage, hypotension and coagulopathy.[1] Please consult the Defitelio SmPC for the full list of all side effects reported with Defitelio.

▼This medicinal product is subject to additional monitoring.

About Defitelio®▼ (defibrotide)
In October 2013, the European Commission granted Marketing Authorisation under exceptional circumstances for Defitelio®▼ (defibrotide) for the treatment of severe hepatic veno-occlusive disease in haematopoietic stem-cell transplantation therapy. It is indicated in patients over one month of age. Defitelio is not indicated in patients with hypersensitivity to defibrotide or any of its excipients or with concomitant use of thrombolytic therapy.

In addition to its existing approved indication in the European Union (EU), defibrotide has the potential to be developed for approval in countries outside the EU and in other indications. Defibrotide has been granted orphan drug designation to treat and prevent VOD by the U.S. Food and Drug Administration (FDA), by the European Medicines Agency (EMA) and by the Korean Ministry of Food and Drug Safety (MFDS), orphan drug designation for the treatment of VOD by the Commonwealth of Australia-Department of Health, and Fast Track designation to treat sVOD by the FDA. Sigma-Tau Pharmaceuticals, Inc. has licensed the rights to commercialise defibrotide for the treatment and prevention of VOD in North America, Central America and South America, subject to receipt of Marketing Authorisation in the applicable territory.

Please consult the Defitelio SmPC before prescribing, particularly in relation to use of medicinal products that increase the risk of haemorrhage, concomitant systemic anticoagulant therapy, medicinal products that affect platelet aggregation, use in patients who have or develop clinically significant acute bleeding requiring blood transfusion, and patients who have haemodynamic instability.

About VOD
Hepatic veno-occlusive disease (VOD) is an early complication in patients undergoing HSCT. In its severe form, VOD can be life-threatening and is associated with multi-organ failure and is fatal in over 80% of patients.[2,3] HSCTs are performed with curative intent in patients with haematological malignancies, selected solid tumours and some non-malignant disorders, such as serious haemoglobinopathies.[4,5] In the EU, VOD is designated as a rare disease, affecting less than five in 10,000 people. Studies have reported a wide range of incidence rates for VOD. Generally, data indicate that approximately 14% of patients undergoing HSCT develop VOD.[3]

About Jazz Pharmaceuticals plc
Jazz Pharmaceuticals plc (Nasdaq: JAZZ) is a specialty biopharmaceutical company focused on improving patients' lives by identifying, developing and commercialising differentiated products that address unmet medical needs. The company has a diverse portfolio of products and/or product candidates in the areas of sleep, haematology/oncology, pain and psychiatry. The company's U.S. marketed products in these areas include: Xyrem® (sodium oxybate) oral solution, Erwinaze® (asparaginase Erwinia chrysanthemi), Prialt® (ziconotide) intrathecal infusion, Versacloz™ (clozapine) oral suspension, FazaClo® (clozapine, USP) HD and FazaClo LD. Jazz Pharmaceuticals also has a number of products marketed outside the United States, including Erwinase® and Defitelio®▼ (defibrotide). For more information, please visit www.jazzpharmaceuticals.com.

About Gentium S.p.A.
Gentium S.p.A., a majority owned indirect subsidiary of Jazz Pharmaceuticals plc, developed Defitelio®▼ (defibrotide) and markets Defitelio in Europe. Gentium is located in Villa Guardia (Como), Italy. For more information, please visit www.gentium.com.

"Safe Harbor" Statement under the Private Securities Litigation Reform Act of 1995
This press release contains forward-looking statements, including, but not limited to, statements related to the therapeutic and commercial potential of Defitelio®▼ (defibrotide), the expected launch of Defitelio in additional European countries and the timing thereof, the potential for defibrotide to be developed for approval in countries outside the EU and in other indications and other statements that are not historical facts. These forward-looking statements are based on Jazz Pharmaceuticals' current expectations and inherently involve significant risks and uncertainties. Actual results and the timing of events could differ materially from those anticipated in such forward-looking statements as a result of these risks and uncertainties, which include, without limitation, risks and uncertainties associated with the company's ability to realise the anticipated revenues from Defitelio; the company's ability to successfully launch and commercialise Defitelio in a timely manner in additional European countries, including possible delays and unforeseen difficulties in obtaining pricing and reimbursement approvals for Defitelio in any of these countries and the possibility that the approved level of governmental pricing and reimbursement for Defitelio in any of these countries may be lower than the company's estimates; the company's ability to successfully manage the risks associated with integrating Defitelio into the company's product portfolio; the company's ability to obtain regulatory approval for defibrotide in other countries, including the United States, and in other indications; and those other risks detailed from time-to-time under the caption "Risk Factors" and elsewhere in Jazz Pharmaceuticals plc's Securities and Exchange Commission filings and reports (Commission File No. 001-33500), including the Annual Report on Form 10-K for the year ending in December 31, 2013 and future filings and reports by the company. Jazz Pharmaceuticals undertakes no duty or obligation to update any forward-looking statements contained in this press release as a result of new information, future events or changes in its expectations.

SOURCE Jazz Pharmaceuticals plc and Gentium S.p.A.

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