PRESS RELEASE

Basilea swaps its isavuconazole North American co-promote rights for full isavuconazole rights outside of North America

Basel, Switzerland, February 28, 2014 – Basilea Pharmaceutica Ltd. (SIX: BSLN) announced today the amendment as of February 27, 2014 of the isavuconazole License, Co-Development and Co-Promotion Agreement with Astellas Pharma Inc. originally executed in 2010, providing Basilea full rights to isavuconazole in all markets outside of the U.S. and Canada in return for Basilea's right to co-promote the product in the U.S. and Canada and its right to receive payments related to co-promotion and EU milestone payments.

Through this agreement Basilea complements its global ceftobiprole rights with rights for isavuconazole in Europe, Asia and the rest of the world outside of the U.S. and Canada, providing Basilea a unique opportunity to optimize the value of two highly complementary and synergistic anti-infective drugs.

Under the terms of the amended agreement, Astellas will remain responsible for the continued development and funding of the isavuconazole global candidemia phase 3 study. Astellas will be responsible for the regulatory filing for the primary treatment of invasive aspergillosis and invasive mucormycosis in the U.S. and Canada and will support preparation of the European filing dossier which Basilea will submit as applicant in Europe. Basilea will continue to be entitled to receive the same regulatory milestone and royalty payments in terms of value from Astellas relating to the U.S. and Canadian territories, with total milestone payments of up to CHF 374 million, including sales milestones. Basilea and Astellas will continue to coordinate their development and manufacturing activities and each company will be responsible for commercial activities in its respective territory. A transition plan for the development, manufacture and commercialization of isavuconazole outside of the U.S. and Canada is currently being developed.

Isavuconazole recently successfully completed the phase 3 non-inferiority SECURE study in invasive aspergillosis showing significant safety advantages. In addition, isavuconazole treated patients with renal impairment and emerging molds such as mucormycosis in the open-label VITAL study, which with the SECURE study data, will form the basis of a regulatory filing in the U.S. and EU.

Ronald Scott, Basilea's Chief Executive Officer, stated: "Through this agreement Basilea secures the full economic benefit of isavuconazole in all markets outside the U.S. and Canada. This provides Basilea and its shareholders with the opportunity for a significant potential economic upside. Isavuconazole complements ceftobiprole which was recently approved in twelve EU member states. We are focusing on the additional opportunities that are now available to Basilea by having rights to two well-differentiated, late-stage hospital anti-infectives. This provides us with exciting new strategic options and opens new opportunities from a franchise perspective. We are reviewing the potential to sell both ceftobiprole and isavuconazole in Europe through distributors or a contract sales organization which could optimize the value of these two synergistic drugs addressing life-threatening infections. We are working with Astellas with a high priority to file isavuconazole initially in the EU and U.S. We also continue to review all options to create the most value for Basilea and its shareholders."

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Financial outlook

Total 2014 operating expenses are estimated to change from CHF 7 to 8 million per month to approximately CHF 8 to 9 million per month. Basilea's 2014 average operating loss is estimated to change from CHF 3 to 4 million per month to approximately CHF 4 to 5 million per month.

About isavuconazole

Isavuconazole (drug substance: isavuconazonium sulfate) is an investigational once-daily intravenous and oral broad-spectrum antifungal for the potential treatment of severe invasive and life-threatening fungal infections. It is currently in phase 3 of clinical development.

Topline results from the randomized, double-blind invasive aspergillosis SECURE phase 3 study showed that isavuconazole was non-inferior to the standard-of-care, voriconazole, as assessed by the primary endpoint of all-cause mortality through day 42. Study drug-related adverse events were significantly lower in the isavuconazole group (42.4%) compared to the voriconazole group (59.8%).

In addition to a potentially improved safety profile, isavuconazole, through its spectrum of activity against molds causing mucormycosis (zygomycosis) and its predictable drug exposure, has the potential to overcome a number of limitations of the current standard-of-care for the treatment of invasive mold infections.

The open-label phase 3 VITAL study enrolled 149 patients. The study included patients with invasive fungal disease caused by emerging fungal pathogens such as Mucormycetes and patients with aspergillosis who had pre-existing renal impairment for which i.v. voriconazole can only be used with caution. VITAL study results show that day 42 all-cause mortality in renally-impaired patients with invasive aspergillosis (n = 20) was 15%. In the SECURE study, which due to the comparator did not allow for enrollment of patients with moderate or severe renal impairment, the mortality rate in patients treated with isavuconazole (n = 258) was 18.6%. In addition, day 42 all-cause mortality in VITAL study patients with confirmed mucormycosis (n = 37), which included patients refractory or intolerant to other antifungal therapies, was 37.8%, which is similar to the mortality rates reported in the literature for the treatment of mucormycosis.

Enrollment in the randomized, double-blind phase 3 isavuconazole study ACTIVE, evaluating the use of isavuconazole i.v. and oral versus caspofungin i.v. followed by oral voriconazole for the treatment of invasive *Candida* infections, is continuing with anticipated completion of enrollment in the first half of 2015.

Isavuconazole demonstrated *in-vitro* and *in-vivo* coverage of a broad range of yeasts (such as *Candida* species) and molds (such as *Aspergillus* species) as well as activity in *in-vitro* studies and in animal models against emerging and often fatal molds including those that cause mucormycosis.

In the U.S., isavuconazole has FDA fast-track status and received QIDP and orphan drug designation for invasive aspergillosis and mucormycosis (zygomycosis). Isavuconazole is being co-developed with Astellas Pharma Inc.

About ceftobiprole

Ceftobiprole is a broad-spectrum intravenous antibiotic from the cephalosporin class for the first-line treatment of severe bacterial infections. It was approved by twelve EU member states for the treatment of hospital-acquired pneumonia (excluding ventilator-associated pneumonia) and community-acquired pneumonia and is currently under regulatory review in Switzerland.

Ceftobiprole is a bactericidal antibiotic. No other single agent has such broad-spectrum activity that includes methicillin-resistant *Staphylococcus aureus* (MRSA) and *Pseudomonas*. It also covers further Gram-positive bacteria such as vancomycin-resistant *Staphylococcus aureus*

(VRSA) and penicillin- and ceftriaxone-resistant *Streptococcus pneumoniae* (PRSP, CRSP) as well as additional Gram-negative pathogens, including Enterobacteriaceae.

Conference call

Basilea Pharmaceutica Ltd. invites you to participate in a conference call on Friday, February 28, 2014, 4 p.m. (CET), during which the company will discuss today's press release.

Dial-in numbers are:
+41 (0) 58 310 50 00 (Europe and ROW)
+1 (1) 631 570 5613 (USA)
+44 (0) 203 059 5862 (UK)

A playback will be available 1 hour after the conference call until Monday, March 3, 2014, 6 p.m. (CET). Participants requesting a digital playback may dial:
+41 (0) 91 612 4330 (Europe and ROW)
+1 (1) 631 982 4566 (USA)
+44 (0) 207 108 6233 (UK)
and will be asked to enter the ID 18124 followed by the # sign.

About Basilea

Basilea Pharmaceutica Ltd. is headquartered in Basel, Switzerland, and listed on the SIX Swiss Exchange (SIX: BSLN). Through the fully integrated research and development operations of its Swiss subsidiary Basilea Pharmaceutica International Ltd., the company focuses on innovative pharmaceutical products in the therapeutic areas of bacterial infections, fungal infections and oncology, targeting the medical challenge of rising resistance and non-response to current treatment options.

Disclaimer

This communication expressly or implicitly contains certain forward-looking statements concerning Basilea Pharmaceutica Ltd. and its business. Such statements involve certain known and unknown risks, uncertainties and other factors, which could cause the actual results, financial condition, performance or achievements of Basilea Pharmaceutica Ltd. to be materially different from any future results, performance or achievements expressed or implied by such forward-looking statements. Basilea Pharmaceutica Ltd. is providing this communication as of this date and does not undertake to update any forward-looking statements contained herein as a result of new information, future events or otherwise.

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