

Gentium S.p.A.

Share Capital Euro 14,956,317

Registered office in Villa Guardia (Como), Piazza XX Settembre no. 2

Registration number with the Register of Enterprises of Como no. 240386 – Tax code and VAT number no. 02098100130

CALL OF THE ORDINARY SHAREHOLDERS' MEETING AND EXTRAORDINARY SHAREHOLDERS' MEETING

The shareholders of Gentium S.p.A. (hereinafter, the “**Company**”) are invited to attend the next ordinary shareholders’ meeting and extraordinary shareholders’ meeting of the Company, to be held, at the time indicated below, with the offices of the Notary Public, Mr. Massimo Caspani, in Via Pessina no. 3, Como, Italy, on April 29, 2011, in first call, and, if necessary, on May 9, 2011, at the same place and time, in second call, in order to discuss and resolve upon the following:

Agenda

Ordinary Shareholders’ Meeting (10:00 a.m. – CET)

1. Approve the 2010 Italian GAAP financial statements of the Company and related documents and allocate the annual operating profits to the Company’s net worth reserve.
2. Set the number of directors of the Board of Directors of the Company and elect members of the Board of Directors of the Company for the 2011/2012 term.
3. Approve director compensation.
4. Approve the engagement of Reconta Ernst & Young S.p.A. as the Company’s independent auditor for fiscal year 2011 with respect to the Company’s U.S. GAAP financial statements and its compensation.

Extraordinary Shareholders’ Meeting (11:00 a.m. – CET)

1. To amend the previous resolution of the Extraordinary Shareholders’ Meeting held on April 27, 2007 and to increase the capital of the Company in cash for a maximum amount of €2,200,000 for the issuance of stock options. To grant the Chairman of the Board of Directors with the power to carry out all the necessary formalities in order to implement the above mentioned resolution.
2. To update and amend Article 6 of the Company’s bylaws in order to confirm the previously granted powers of the Board of Directors, to increase the capital of the Company, issue convertible bonds and, in each case, exclude or limit the option right of the shareholders if it is in the interest of the Company. To grant the Chairman of the Board of Directors with the power to carry out all the necessary formalities in order to implement the above mentioned resolution.

The shareholders may attend the Shareholders' Meetings if they are provided with the relevant authentications by the authorized intermediary, pursuant to article 85 of the Legislative Decree no. 58 of 1998 and article 34 of the CONSOB resolution no. 11768 of December 23, 1998. Holders of the Company's American Depositary Shares ("ADSs") of record on March 2, 2011 will be able to instruct The Bank of New York, the Company's depository of the ordinary shares representing the ADSs, to vote those ordinary shares at the meeting pursuant to the terms of the Deposit Agreement dated as of June 15, 2005 between the Company and The Bank of New York.

The documents relating to the Shareholders' Meetings will be deposited at the registered office of the Company according to the law.

Villa Guardia (Como), March 2, 2011

The Chairperson of the Board of Directors

(Dr. Khalid Islam)