



## Press Release

### Shire enhances its orphan drug pipeline with the acquisition of a new clinical candidate for Metachromatic Leukodystrophy

**Basingstoke, UK and Cambridge, MA, US – 24 April, 2008** – Shire plc (LSE: SHP, NASDAQ: SHPGY), the global specialty biopharmaceutical company, announces the acquisition of arylsulfatase –A (ASA) an Enzyme Replacement Therapy (ERT) in Phase 1-2 clinical trials for Metachromatic Leukodystrophy (MLD) from the Danish company Zymenex A/S (Zymenex).

MLD is a serious, life-limiting disease in which patients experience progressive irreversible neurological damage. MLD is caused by a deficiency in the enzyme ASA which causes an excess concentration of sulphatide in cells and an ensuing breakdown of myelin. There are approximately 2,000 MLD patients in developed world markets<sup>1</sup>.

The newly acquired ASA product, currently known as METAZYM™, has completed a Phase Ib clinical trial in 12 MLD patients in Europe and an extension to this study is ongoing. The product has received Food and Drug Administration (FDA) approval for its Investigational New Drug (IND) application to initiate a phase 2 clinical trial and has been granted Orphan Drug Designation in the United States and in the European Union.

Sylvie Grégoire, President of Shire's Human Genetic Therapies business, commented: "This product fits very well with Shire's ERT portfolio of treatments for Lysosomal Storage Disorders (LSD). Shire HGT has been committed to MLD and by acquiring this mid-stage clinical program we hope to bring a MLD treatment to patients two years earlier than anticipated."

Shire is making a payment to Zymenex of US\$135 million for the acquisition of global rights to the product upon completion of the transaction, which is conditional upon the receipt of customary consents. Zymenex is also providing certain transition services, including know-how transfer, for up to six months after completion. The transaction includes no royalties or other downstream payment obligations.

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<sup>1</sup> Scriver et al 1995

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## Notes to editors

### **Disease Background**

Metachromatic Leukodystrophy (MLD) is in the family of lysosomal storage diseases (LSD's). MLD is an autosomal recessive disease caused by a deficiency of the lysosomal enzyme arylsulfatase A (ASA) which results in an increased concentration of sulphatide in cells of the brains and in non-neural tissue, such as the kidneys and gallbladder. When these sulfated glycolipids accumulate in the brain, they cause a breakdown of myelin, a substance that protects the nerves in the brains and in the rest of the body. This breakdown is what makes MLD a progressive, neurodegenerative disease.

### **Symptoms and Patient Outlook**

Sulfatide accumulation in the central and peripheral nervous system leads to destruction of the myelin sheath (demyelination). MLD has a spectrum of disease which can arise in infants and young children with a range of symptoms, though most are related to motor and cognitive decline. Most MLD patients are normal at birth but often die before age 20, with some patients within the first few years of life. During the final stages all patients reach a decerebrate, vegetative state. The majority of cases are late infantile or juvenile patients. The number of adult patients exhibiting mild forms of MLD is unknown, as adult onset MLD can present with symptoms similar to psychosis.

## **SHIRE PLC**

Shire's strategic goal is to become the leading specialty biopharmaceutical company that focuses on meeting the needs of the specialist physician. Shire focuses its business on attention deficit and hyperactivity disorder (ADHD), human genetic therapies (HGT), gastrointestinal (GI) and renal diseases. The structure is sufficiently flexible to allow Shire to target new therapeutic areas to the extent opportunities arise through acquisitions. Shire's in-licensing, merger and acquisition efforts are focused on products in niche markets with strong intellectual property protection either in the US or Europe. Shire believes that a carefully selected portfolio of products with strategically aligned and relatively small-scale sales forces will deliver strong results.

For further information on Shire, please visit the Company's website: [www.shire.com](http://www.shire.com).

## **"SAFE HARBOR" STATEMENT UNDER THE PRIVATE SECURITIES LITIGATION REFORM ACT OF 1995**

Statements included herein that are not historical facts are forward-looking statements. Such forward-looking statements involve a number of risks and uncertainties and are subject to change at any time. In the event such risks or uncertainties materialize, Shire's results could be materially affected. The risks and uncertainties include, but are not limited to, risks associated with: the inherent uncertainty of pharmaceutical research, product development including, but not limited to, the successful development of JUVISTA<sup>®</sup> (Human TGFβ3) and veleglucerase alfa (GA-GCB); manufacturing and commercialization including, but not limited to, the establishment in the market of VYVANSE<sup>™</sup> (lisdexamfetamine dimesylate) (Attention Deficit and Hyperactivity Disorder ("ADHD")); the impact of competitive products, including, but not limited to, the impact of those on Shire's ADHD franchise; patents, including but not limited to, legal challenges relating to Shire's ADHD franchise; government regulation and approval, including but not limited to the expected product approval date of INTUNIV<sup>™</sup> (guanfacine extended release) (ADHD); Shire's ability to secure new products for commercialization and/or development; and other risks and uncertainties detailed from time to time in Shire plc's filings with the Securities and Exchange Commission, including Shire plc's Annual Report on Form 10-K for the year ended December 31, 2007.