Pharming Group NV

(NYSE Euronext: PHARM)

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HEALTH ■ SCIENCE ■ PRODUCTS

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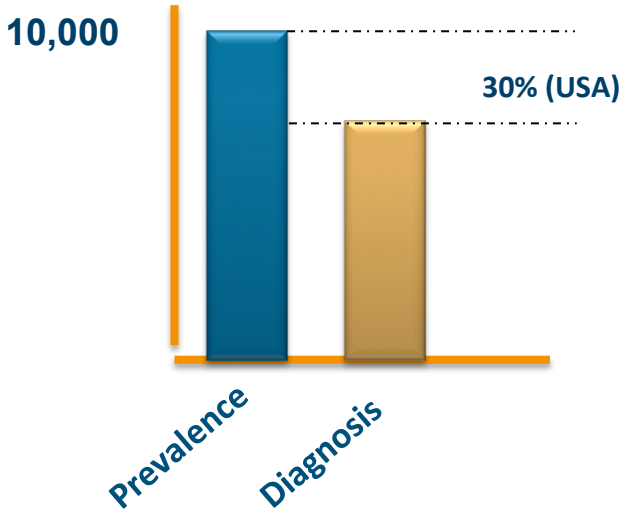
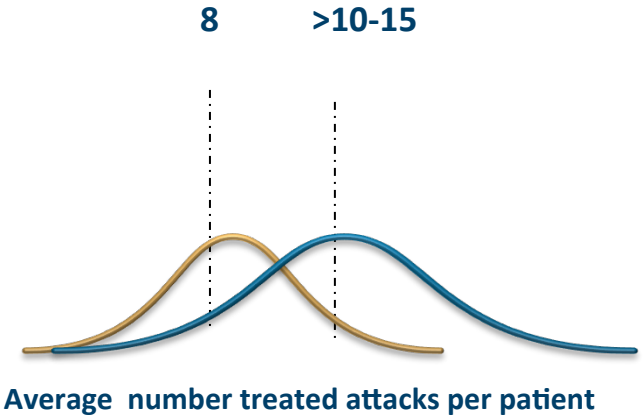
Highlights

- **PDUFA date for lead product Ruconest® (rhC1INH) in acute treatment of attacks of Hereditary Angio- Edema (HAE): 16 July 2014**
- **US partner: Santarus/ Salix Pharmaceuticals (SLXP)**
  - Differentiated competitive profile in US market **– potential to be best-in-class**
  - Preparations for US launch are ongoing (US$20M milestone at first commercial sale)
- **Significant potential from additional indications**
  - Prophylaxis of HAE: RCT to start 2H2014
  - Acute Pancreatitis: Seeking FDA guidance
- **Ruconest® rolling out in Europe for acute HAE (European partner: Sobi)**
  - In market sales increasing; positive feed- back patients and prescribers
- **Pipeline development: Strategic product development collaboration with Shanghai Institute for Pharmaceutical Industry (SIPI) a Sinopharm Company**
  - New product development up to IND and manufacturing at SIPI

Hereditary Angio-Edema (HAE)

- Rare genetic disorder caused by mutations in the gene encoding C1 esterase inhibitor (C1INH)
  - Patients present with swelling, severe abdominal pain, or acute airway obstruction





- Prevalence estimates range from 1 in 10K–50K
- 75% of patients present before age 15
- Misdiagnosis is common; rate of diagnosis varies widely (US is most developed market)

- Increase in attacks treated per patient per year
  - Still significant long term steroid prophylaxis
    - Ineffective and high liver toxicity risk
  - Laryngeal attacks are potentially lethal
  - More aggressive treatment of all type of attacks

Inhibition of C1 esterase is Gold Standard for HAE treatment (protein replacement)

Ruconest commercialisation

- **EU rollout continuing by partner Sobi**
  - Improving in- market sales under challenging EU market access conditions
    - 1Q2014 revenues from sales to Sobi €0.9M
  - Significant market penetration in several Central/ Eastern European markets
  - Consistent and significant repeat use and high patient and physician satisfaction
- **Approved in Israel/ final stage of regulatory review in Turkey**
  - Partnered with Megapharm (Israel) and Eczasibasi (Turkey)
  - SE- Asian territories partnered with Transmedic Pte. and Hyupjin Corp.
  - China, Taiwan, Hong Kong and Macau partnered with SIPI
- **During 2014; (ex- US) sales to partners to increase by >€2 M to €3M**
- **Unlimited supply capabilities and significant economies of scale**
  - **Rapidly scale- able supply chain**
  - **Technology transfer to SIPI to set- up future second supply source in Shanghai**

US market: Rapid growth, significant potential

- **HAE disease awareness in the US continues to improve, leading to more patient identification***
- **FY 2013 sales for acute treatment increased to approx. US$ 275M from US$ 156M for FY2012 (50% growth) excluding Berinert® sales (not disclosed***)**
  - US$ 235M Firazyr® (US$75M in Q12014)**
  - US$ 40.5M Kalbitor® (US$12.5M in Q12014)**
  - Treatment costs estimated at US$70k/ annum***
- **FY 2013 sales for prophylaxis (Cinryze®) increased to approx. US$395M from US$327M for FY 2012; First 2 months 2014 US$86M****
- **More patients seeking treatment for moderate symptoms***
  - Guidelines recommend treating all attacks since any one could become severe
  - Many patients use multiple products, patient driven therapies
  - Significant steroid usage remains to date

* Leerink Swann, competitor interviews, 13 Sept13, 2012,

** Quarterly results 2014 and FY 2013 results SEC filings DYAX, SHPG

*** Seeking alpha an overview of HAE 18 Sep 2012

HAE treatment options (published data)

		recombinant C1 Inhibitor	plasma derived C1 Inhibitor		bradykinin receptor antagonist	kallikrein inhibitor
		Ruconest^	**Cinryze^^**	**Berinert**	**Firazyr****	**Kalbitor^^^**
Efficacy		**Excellent**	**Good**	**Good**	**Good**	**Good**
	Dosing (C1INH)	**50 U/kg***	**~ 12 U/kg**	**20 U/kg**		
	Treatment type	**Any acute**	**Prophylaxis**	**Limited******	**Any acute**	**Any acute**
	Response < 4h	**80-100%**	**~ 60%**	**70%**	**58-74%**	**73%**
Safety concerns		**Very low risk of allergic reaction**	**Warning: Risk of blood clots**	**Warning: Risk of blood clots**	**97% injection site reactions**	**Black box warning 3.9% anaphylaxis**
	Plasma risk	**NO**	**YES**	**YES**	**No**	**No**
Purity (C1INH)		**>99.9%**	**±80%**	**±95%**		
Relapse / worsening		Uncommon	Uncommon	Uncommon	**11-31%*****	**21%**
Administration		IV	IV	IV	SQ	SQ (no self-administration)

***Optimal efficacy of C1INH therapy is achieved at doses ≥50 U/kg (**"Target levels of functional C1-inhibitor in hereditary Angioedema". Allergy, C. E. Hack, A. Relan, E. S. van Amersfoort & M. Cicardi**)**

**Icatibant Clinical Briefing Document, CDER, FDA, 2011./ Aberer, et al. Ann Allergy Asthma Immunol 2010; 105(5):P238

** *Cicardi et al, N Engl J Med 2010;363:532-41.; Aberer, et al. Ann Allergy Asthma Immunol 2010; 105(5):P238; Lumry, et al. Ann Allergy Asthma Immunol. 2011;107:529 –537.

****Berinert not licensed for peripheral attacks in the US,
^Ruconest approved in EU and Israel, ^^Cinryze not licensed for acute therapy in US. ^^^Kalbitor not approved in EU.

Technology (protein expression and production) Platform

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Pharming protein expression and production platform (rabbit milk)

- Low set- up costs compared to cell based systems
- No changes in yield/ protein structure from up- scaling
- Capable of yielding high concentrations of complex (highly glycosylated) proteins (expression of rhC1INH at ≅12 grams/ liter) in combination with significant volumes (150-200 ml/day)
- Minimal differences with human glycosylation pattern
- Flexible supply chain and reduced financial risk/ exposure: Low cost intermediate holding stage (frozen milk) and rapid scale-ability
- Wide applicability, significant experience
  - Plasma proteins (C1INH/ F VII/ F VIII/ F IX
  - Metabolic enzymes (α- glucosidase)
  - Monoclonal antibodies
  - Hormones

Strategic product development collaboration

SIPI (Shanghai Institute for Pharmaceutical Industry: A Sinopharm company)

- Product development at SIPI
  - Under Pharming's fully ICH compliant QA systems
  - Compliant with CFDA, FDA and EMA standards
  - Funded by SIPI up to IND
  - Aligned clinical development (SIPI funds China/ Pharming funds ROW)

- Technology transfer of Pharming platform to SIPI facilities in Shanghai
  - Initial projects C-1 Inhibitor and Factor VIII
  - Includes manufacturing of (future) finished products

- SIPI's product development resources and SIPI's favourable cost structures for development and manufacturing combined with the competitive features of the platform

SIPI collaboration

- **Commercialisation rights: SIPI China/ Pharming ROW**
  - Reciprocal royalties at 4%: SIPI (China)/ Pharming (ROW)
  - SIPI to pay product related milestones for all future products developed
  - SIPI to supply Pharming on “cost plus” basis for ROW

- SIPI pays €1.26 million upfront and € 0.84 million technology transfer fees and all Pharming technology transfer related expenses

- SIPI pays €0.3 million at receipt of Ruconest drug importation license
  - Until completion of technology transfer, Pharming to supply SIPI with Ruconest as imported product (“cost plus” basis and 4% royalties)

Financial and investment highlights

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Financials

- Stabilised balance sheet and lean cost structure
  - 30 April 2014 cash balance of €30M/ Q1 OPEX €3.5M
  - Significant Ruconest inventories available
  - Increasing revenues from sales (Ex-US)

– **Subject to US Approval (PDUFA date 16 July 2014)**
  – First US commercial sale triggers US$20M milestone
  – Proceeds from supply of Ruconest to SLXP
    - 30%- 40% of SLXP Ruconest net sales
      – Up to US$45 million in sales related milestones

- Investment in expansion of rhC1INH franchise
  - Prophylaxis of HAE: RCT to start 2H2014
- Collaborative leveraging of potential of the platform
  - C1 inhibitor technology transfer to SIPI
  - Factor VIII development at SIPI

Investment Highlights

- **PDUFA date Ruconest® for acute HAE 16 July 2014.**
  - Differentiated competitive profile/ Rapidly expanding US acute market segment estimated at >US$ 400M + per annum
  - Significant potential near term milestone US$ 20M (first US commercial sale)
  - Revenues from US net sales between 30-40%
- **Significant up- side potential from additional indications**
  - Prophylaxis of HAE and Acute Pancreatitis
- **Ruconest® sales increasing in Europe and ROW**
- **Pipeline development**
  - New product development at SIPI and supply by SIPI
- **Stabilised balance sheet + low operating costs: Basis for future profitability**
  - Increasing ROW sales and US market entry to drive economies of scale/ reduction of COGS
  - Significant value inflexion points ahead

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