Pharming Group NV

(NYSE Euronext: PHARM)

Sijmen de Vries, MD, MBA
Chief Executive Officer

Annual General Meeting of Shareholders
Leiden, 15 May 2013

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HEALTH ■ SCIENCE ■ PRODUCTS

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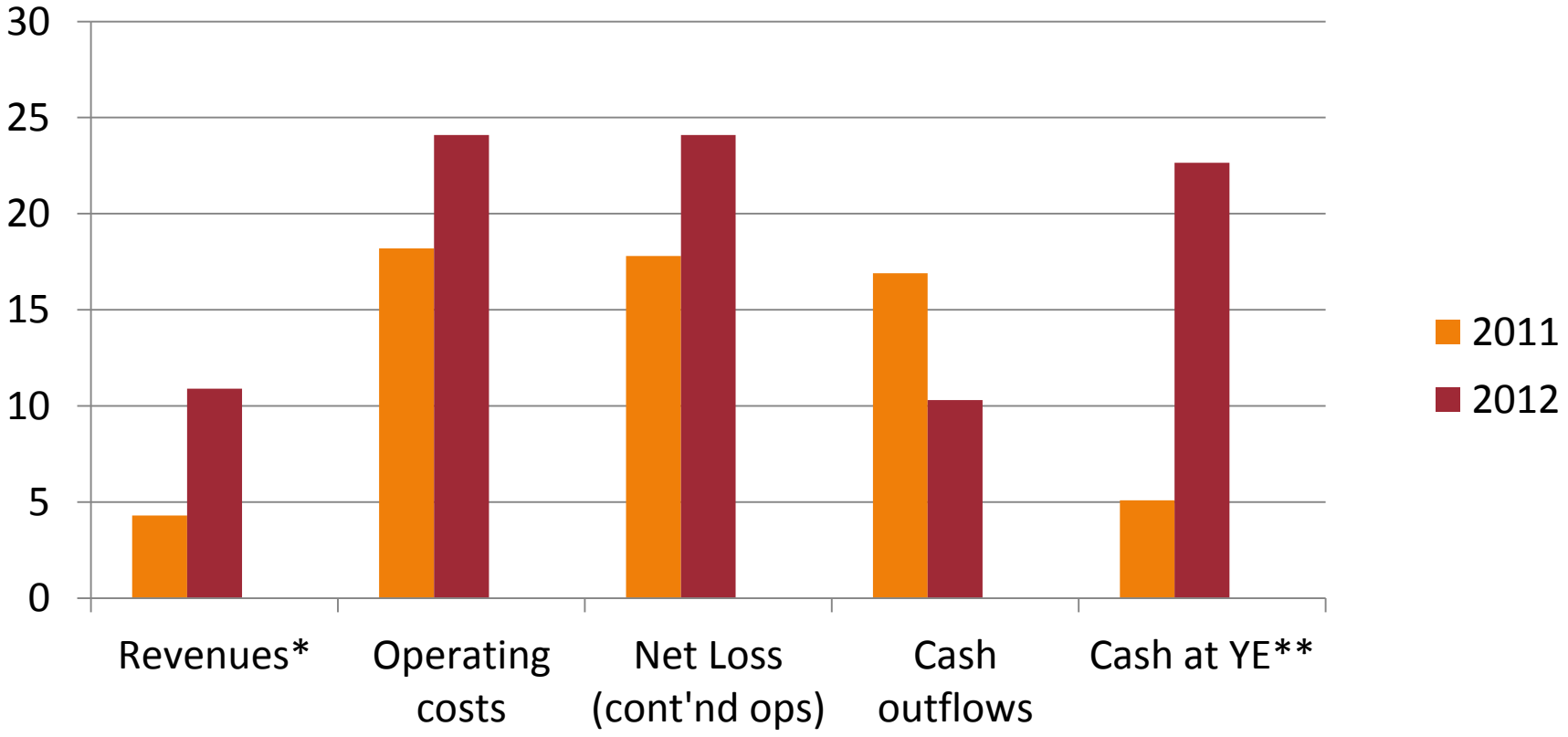
Operations 2012

- **Validation and production runs completed and certification process initiated with EMA for 2nd downstream production facility at Sanofi- Chimie**
  - Up- scaling of production capacity and reduction in cost of goods
  - Process and facility approved by EMA in March 2013
- **Significant downsizing of organisation initiated, to be completed by the end of 1H2013**
  - Termination of cattle based research, closing and sale of US research facilities
  - Significant downsizing of Dutch operations
  - Total headcount reduction from 69 FTE in 2H2012 to approx 35 FTE in 2H2013
- **Study 1310 (Phase III US pivotal study) was successfully completed in early November and a US$ 10 M milestone payment from SNTS was received**
- **Ruconest exploratory (Phase II) open label study in prophylaxis of HAE and additional supporting evidence of absence of pro- thrombotic effects when treating patients with HAE were published in international peer reviewed journals**

Ruconest commercialisation 2012

- **EU rollout continuing**
  - Challenging EU market access conditions (FY2012 sales € 0.9 M)
  - Reimbursement discussions at national, regional and local levels ongoing (Italy, Spain)
  - Sobi recently re- aligned resources to rapidly adapt to leverage reimbursement approvals
  - Steadily increasing numbers of patients with consistently “repeat Rx”
- **Increased geographical coverage**
  - Regulatory review ongoing in Turkey, Israel, and the SE- Asian territories partnered with Transmedic Pte. and Hyupjin Corp.
- **Partnering discussions in progress**
  - China, Taiwan, Macau and Hong- Kong
  - Latin American territories
- **Unlimited supply capabilities and significant economies of scale**

Financials 2012



* Includes €7.9M SNTS milestone payment

** ”Pro-forma” YE 2012, includes €16.35M January 2013 CB financing

Dutch Corporate Governance Code

- Corporate Governance Statement 2013 on website www.pharming.com/corporate

- Pharming complies with the Dutch Corporate Govenance Code except for sections: (unchanged vs. 2011)
  - II.2.4. (Options for the management board)
  - III.3.1. (Profile of the Board of Supervisory Directors)
  - III.6.5 (Ownership and transactions in securities other than issued by the Company)
  - IV.3.1 (Follow meetings in real time)
  - IV.3.12 (Independent third party to hold proxies)
  - IV.3.13 (Outline policy in bilateral contact with shareholders)
  - III.5.4c-III.5.4d and V.3.1.-V3.3. (Internal auditor)

Perspectives for 2013 and beyond

- **BLA Ruconest® for acute HAE sumitted 17 April 2013.**
  - Standard 12 month (PDUFA V) review
  - Rapidly expanding US acute market segment at US$ 156M + per annum
  - Differentiated competitive profile in US market; potential for treatment paradigm shift
  - Significant potential near term milestones: US$ 5M (acceptance of BLA) US$ 20M (first US commercial sale) from US partner Santarus (SNTS)
- **Significant up- side potential from additional indications**
  - USA: Prophylaxis of HAE and Acute Pancreatitis; seeking further FDA guidance
  - Ischemia Reperfusion Injury related indications (on-going, pre-clinical)
- **Ruconest® (rhC1INH) rolling out in Europe (European partner: Sobi)**
  - Sobi has recently re-aligned commercialisation resources
- **Protein manufacturing technology platform poised to replicate rhC1INH success**
- **Downsized infrastructure; scope for reduction in operating costs**
- **Balance sheet: Cash runway covers standard US regulatory review**
  - Significant value inflexion points ahead

U.S. Market opportunity

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US market dynamics: growing demand

- **HAE disease awareness in the US continues to improve, leading to more patient identification***
- **FY 2012 sales increased from US$ 56M to US$ 156M (acute HAE segment) (179% growth) (Berinert® sales not disclosed***)**
  - US$ 116M Firazyr® (US$ 33M; 2011)**
  - US$ 40M Kalbitor® (US$ 23M;2011)**
  - Treatment costs estimated at US$70k/ annum***
- **More patients starting to seek treatment for moderate symptoms***
  - Guidelines recommend treating all attacks since any one could become severe
  - Many patients use multiple products
- **Hereditary nature of HAE results in newly diagnosed patients and their relatives being particularly strong source of Rxs***
  - New patients tend to be very motivated early on or after seeing family members with the disease

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

** FY 2012 10-Q filings VPHM, DYAX, SHPG

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

U.S. Competitor profiles

		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		
	Type of attack	Any	Any	Limited	Any	Any
	Response < 4h	80-100%	60%	70%	58-74%	73%
Safety concerns		Very low risk of allergic reaction	Low risk of blood clots	Low risk of blood clots	97% injection site reactions	Black box warning 3.9% anaphylaxis
	Plasma risk	No	YES	YES	No	No
Purity (C1INH)		99%	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**)**

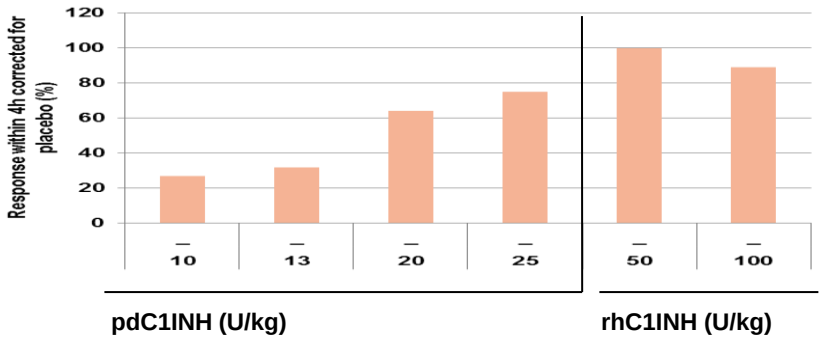
**Icatibant Clinical Briefing Document, CDER, FDA, 2011./ Aberer, et al. Ann Allergy Asthma Immunol 2010; 105(5):P238

** *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.

Ruconest approved in EU only, Cinryze not licensed for acute therapy in US.
Kalbitor not licensed in EU.

Ruconest: potential to be best in class

- **C1 esterase inhibition is Gold Standard for HAE treatment (protein replacement)**
- **Best-in-class efficacy and cleanest safety profile (confirmed in Study 1310)**
  - Absence of thrombo- embolic complications, no anaphylactic reactions
- **Ability to administer higher dosing and clean safety profile provides significant efficacy and safety advantages over plasma derived C1 Inhibitor**
  - Optimal efficacy of C1INH therapy is achieved at doses ≥50 U/kg*



- **Ruconest profile could provide some (US) patients with the first dependable option to dose "acutely" for all types of attacks instead of having to rely on (expensive) and only partly efficacious** prophylactic therapy**

*Allergy, 2011 C. E. Hack, A. Relan, E. S. van Amersfoort & M. Cicardi)
** N Engl J Med 2010;363:513-22, Zuraw et al

Technology Platform

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Validated transgenic rabbit (milk) platform

Value creation by partnerships

Low set up cost, low risk up-scaling, low cost intermediate bulk (frozen milk) holding stage

IP and know-how protection

- US patent (2027)

Relevant for most therapeutic proteins

Complex proteins of high quality with relatively high yields

Initial focus on blood clotting factors & metabolic enzymes

High yields drive potential for low cost of manufacturing

Often achieving significantly higher expression (1-15 g/L) of recombinant human proteins compared to cell based systems (150 fold with rhC1Inhibitor)

Successful expression of recombinant human proteins in milk of transgenic animals achieved by Pharming and others

Plasma Proteins	• **Serpins: C1 Inhibitor, α1-antitrypsin and Antithrombin-III** • **Clotting Factors VII, VIII, IX and von Willebrand Factor** • **Albumin and Fibrinogen**
Metabolic enzymes	• **α–Glucosidase**
Monoclonal antibodies	• **High expression levels in various species: up to 30 g/L**
Hormones	• Human Growth Hormone • Follicle Stimulating Hormone
Structural proteins	• Collagen
Others	• Protein vaccines • Lactoferrin and Lysozyme • Bile-salt stimulated lipase

Outlook 2013 and beyond

- **Ruconest BLA submitted to US FDA (April 2013)**
  - Acceptance for review by FDA triggers US$5M milestone
  - First US commercial sale triggers US$20M milestone
  - Proceeds from US commercialisation by SNTS
    - 30%- 40% of Ruconest net sales
    - Up to US$45 million in sales related milestones

- **Expansion of rhC1INH franchise**
  - Prophylaxis of HAE
  - Acute pancreatitis
  - Ischemia Reperfusion Injury

- **Leverage potential of the platform**
  - New proteins and partnerships

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