Diverse info over Pharming

Pharming
www.pharming.com

Statuten Pharming
Articles of Association Pharming Group N.V.

Een reis in de tijd
https://www.iex.nl/Forum/Topic/1345957/Pharming_Een-korte-chronologische-reis-in-de-tijd.aspx?page=2#13535041

Pharming Facts
https://www.iex.nl/Forum/Topic/1334048/Pharming_Pharming-Facts-II.aspx?page=10#13533531

Diverse info
https://nl.marketscreener.com/koers/aandeel/PHARMING-GROUP-N-V-12738425/koersen/

Jaarstukken 2021
https://www.pharming.com/nl/node/304

Diverse bedrijfsgegevens
https://beurs.fd.nl/noteringen/96535/pharming/algemeen

Historisch koersoverzicht
https://www.iex.nl/Forum/Topic/1291122/Pharming_Historisch-koersoverzicht-van-Pharming.aspx?page=25#13564626

Historisch aandelenoverzicht
https://www.iex.nl/Forum/Topic/1290314/Pharming_Historisch-aandelen-overzicht-van-Pharming.aspx?page=2#13433986

Eigenaarschap aandelen
https://www.morningstar.com/stocks/xams/pharm/ownership

Filmpje HAE voor beginners
https://www.youtube.com/watch?app=desktop&v=9a1EE31Zkg4&feature=emb_title

Over Ruconest
https://www.ema.europa.eu/en/medicines/human/EPAR/ruconest

Productkenmerken Ruconest
https://www.ema.europa.eu/en/documents/product-information/ruconest-epar-product-information_nl.pdf

Prijs Ruconest
https://www.farmacotherapeutischkompas.nl/bladeren/preparaatteksten/c/conestat_alfa

Beloningsbeleid
https://usir.pharming.com/static-files/ee4b578b-66a9-4a07-8566-b5bc1da432e5

Overview Total Remuneration CEO

The following table sets out the total remuneration for the Executive Director/CEO (and former members of the board of management), including the awards and pay-outs (i.e., in 2022 for the short-term variable amount) based on the outcome of the performance assessment for 2021, as described in the preceding section.

in US$ '000	Fixed remuneration	Short term variable: annual bonus	Share based payments	Post-employment benefits	Other	TOTAL
Mr Sijmen de Vries, CEO and Executive Director	2021: 681 (32%) 2020: 614 (21%) 2019: 568 (36%) 2018: 579 (36%) 2017: 537 (33%)	2021: 357 (17%) 2020: 431 (15%) 2019: 347 (22%) 2018: 506 (32%) 2017: 373 (23%)	2021: 1,264 (44%) 2020: 1,739 (59%) 2019: 546 (35%) 2018: 384 (24%) 2017: 606 (37%)	2021: 120 (6%) 2020: 107 (4%) 2019: 81 (5%) 2018: 96 (6%) 2017: 89 (5%)	2021: 38 (2%) 2020: 37 (1%) 2019: 36 (2%) 2018: 38 (2%) 2017: 36 (2%)	2021: 2,460 2020: 2,927 2019: 1,578 2018: 1,603 2017: 1,641
Mr. Bruno Giannetti, CMO	2021: - 2020: 402 (28%) 2019: 371 (38%) 2018: 378 (38%) 2017: 349 (34%)	2021: - 2020: 201 (14%) 2019: 190 (20%) 2018: 275 (28%) 2017: 210 (20%)	2021: - 2020: 708 (50%) 2019: 324 (33%) 2018: 238 (24%) 2017: 371 (36%)	2021: - 2020: 85 (6%) 2019: 78 (8%) 2018: 91 (9%) 2017: 88 (8%)	2021: - 2020: 27 (2%) 2019: 9 (1%) 2018: 9 (1%) 2017: 17 (2%)	2021: - 2020: 1,424 2019: 973 2018: 992 2017: 1,036
Mr. Robin Wright, CFO	2021: - 2020: 155 (24%) 2019: 255 (53%) 2018: 362 (47%) 2017: 335 (45%)	2021: - 2020: 14 (2%) 2019: 167 (25%) 2018: 175 (23%) 2017: 153 (20%)	2021: - 2020: 107 (17%) 2019: 128 (18%) 2018: 197 (25%) 2017: 229 (30%)	2021: - 2020: 15 (2%) 2019: 26 (4%) 2018: 40 (5%) 2017: 38 (5%)	2021: - 2020: 350 (55%) 2019: - 2018: - 2017: -	2021: - 2020: 641 2019: 676 2018: 774 2017: 755

The remuneration amounts paid in 2021 to the Executive Officers are not required to be disclosed according to Dutch law and accordingly are not disclosed herein.

Interview met Sijmen de Vries in het Leidsch Dagblad van 23 februari 2022, o.a. over omzetverwachting leniolisib

De lange adem van Pharming: beursgenoteerd biotechbedrijf veranderde van brekebeentje in winstmachine | De koers van het Bio Science Park | Leidsch Dagblad

Intellectuele eigendommen Pharming
https://uspto.report/company/Pharming-Intellectual-Property-B-V

Over Ruconest uit koemelk
13549180.pdf (belegger.nl)

Over het ‘complementsysteem
https://www.ncbi.nlm.nih.gov/books/NBK27100/
https://link.springer.com/article/10.1007/s10875-021-00972-1

Overname commerciële rechten SOBI
https://www.pharming.com/nl/node/172

Europees regulatoir system geneesmiddelen
https://www.ema.europa.eu/en/documents/leaflet/european-regulatory-system-medicines-european-medicines-agency-consistent-approach-medicines_nl.pdf

https://www.ema.europa.eu/en/documents/other/laboratory-patient-journey-centrally-authorised-medicine_nl.pdf

Advies First Berlin
Geen link beschikbaar. Op verzoek kan ik het document splitsen en in delen posten (tenzij iemand de link al heeft).

De presentatie van dr. M. Osthoff van 29 juni 2021
Geen link beschikbaar. Te omvangrijk om het document hier mee te nemen. Kan ik op verzoek beschikbaar stellen.

Studies/trials waarbij Ruconest wordt ingezet of Pharming is betrokken

Naam trial	Fase	Start*	PCD*	SCD*	Link*
Preventie infectie bij COVID-patiënten (CH/Br/Mex)	2	6 aug 2020	15 sep 2021	15 sep 2021	https://clinicaltrials.gov/ct2/show/NCT04414631 https://data.snf.ch/covid-19/snsf/198403
Preventie infectie bij COVID-patiënten (VS)	2	30 nov 2020	15 aug 2021	30 nov 2021	https://clinicaltrials.gov/ct2/show/NCT04530136
Evaluatie verbetering neurologische symptomen na COVID	4	30 dec 2020	jan 2022	jan 2022	https://www.clinicaltrials.gov/ct2/show/NCT04705831
Nierfalen (AKI)	2	21 apr 2021	dec 2022	dec 2023	https://clinicaltrials.gov/ct2/show/NCT04912141?cond=ruconest&draw=3&rank=17 Verdieping trial: https://www.jacc.org/doi/pdf/10.1016/j.jcin.2019.11.021
APDS (leniolisib)	2/3	24 aug 2015	16 aug 2021	16 aug 2021	https://clinicaltrials.gov/ct2/show/NCT02435173?term=Leniolisib&draw=2&rank=1
APDS – extension (leniolisib)	2/3	8 sep 2016	1 sep 2026	1 sep 2026	https://clinicaltrials.gov/ct2/show/NCT02859727?term=Leniolisib&draw=2&rank=2
DGF	1	21 jun 2019	jan 2022	jan 2022	https://clinicaltrials.gov/ct2/show/NCT03791476?term=Ruconest&cond=Nephropathy&draw=2&rank=2
Pre-eclampsia	1/2	?	?	?	https://www.clinicaltrialsregister.eu/ctr-search/trial/2018-002904-14/NL
Hemofilie A (rhFVIII)	pre-kl.	nnb.	nnb.	nnb.	https://adisinsight.springer.com/drugs/800043667
PAIR-TAVI	2	16 mrt 2022	jan 2024	jan 2024	https://clinicaltrials.gov/ct2/show/NCT05145283
Werking en bijwerking plasma-C1Inh en rcC1Inh	obs. studie	jul 2011	jun 2021	dec 2021	https://clinicaltrials.gov/ct2/show/NCT01397864?term=conestat+alfa&recrs=ab&draw=2&rank=4
Evaluatie veiligheid Ruconest	obs. studie	16 jul 2018	16 jul 2021	16 aug 2021	https://clinicaltrials.gov/ct2/show/NCT03697187?term=conestat+alfa&recrs=ab&draw=2&rank=1

Honden met IMHA	Tussen april en oktober publicatie in ‘Cornell Dog Watch’ en ‘The Bark’. Tevens presentatie tijdens een wetenschappelijk congres.	https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6937664/

* rood : afgebroken studie
* Start : actual study start date
* PCD : estimated primary completion date
* SCD : estimatedstudy completion date
* CT.gov : deadlines updates trials

Overzicht DM van concurrerende markt

Acuut

B2 Receptor
- Firazyr (Takeda/Shire) – Bradykinin receptor antagonist - Subcutaan (FDA & EMA)

C1-INH
- Ruconest (Pharming) – C1-INH - Intraveneus (FDA & EMA)
- Berinert (CSL) – C1-INH - Intraveneus (FDA & EMA)
- Cinryze (Takeda/Shire) – C1-INH - Intraveneus (EMA)

Plasma kallikrein
- Kalbitor (Takeda/Shire) – Kallikrein inhibitor - Subcutaan (FDA & EMA)

Profylaxe

B2 Receptor
Nog geen

C1-INH
- Cinryze (Takeda/Shire) – C1-INH - Intraveneus (FDA & EMA)
- Haegarda (CSL) – C1-INH - Subcutaan (FDA & EMA & Canada onlangs)

Plasma kallikrein
- Takhzyro/Lanadelumab (Takeda/Shire) - Kallikrein inhibitor - Subcutaan (FDA & EMA)
- Orladeyo (Biocryst) - Oral capsule (US, China, Japan, VK, EMA)

In ontwikkeling

- PHVS416 (Pharvaris) – Acuut
  - Orale toediening, klein molekuul en daardoor snel werkzaam (15-30min).

- Gebaseerd op het al jaren bewezen Icatibant
- Versneld traject, afgeleide van de profylaxe toediening
- Phase II gestart, eerste patiënt behandeld feb 2021, resultaten 2022
- Studie uitgebreid naar de US

- PHVS416 (Pharvaris) – Profylaxe
  - Orale toediening, klein molekuul en daardoor snel werkzaam (15-30min).
  - Gebaseerd op het al jaren bewezen Icatibant
  - 1 a 2 maal daags toedienen
  - Phase II start in 2021, resultaten 2022
  - FDA Acceptance IND App april 2021
  - Open b2-receptor markt

- PHVS719 (Pharvaris) – Profylaxe
  - Orale toediening, klein molekuul en daardoor snel werkzaam (15-30min).
  - Gebaseerd op het al jaren bewezen Icatibant
  - extended release form. PHA121 voor profylaxe gebruik
  - Phase I start in 2021, resultaten 2022

- KVD900 (Kalvista) - Acuut
  - Orale toediening, kallikrein inhibitor.
  - Eerste Phase II data gepubliceerd
  - Today's data show that KVD900 halts HAE attack progression and also provides rapid relief by shortening the time to symptom resolution
  - Werving patiënten fase III lopend
  - Fase III data voorzien half 2023 voor NDA filing te onderbouwen

- KVD824 (Kalvista) - Profylaxe
  - Werving patiënten fase II lopend
  - Fase II data voorzien mid 2023
  - Tweemaal daags orale toediening voor profylaxe

- Oral Factor XIIa inhibitor (Kalvista) - Profylaxe
  IND 2023

- Donidalorsen (eerst IONIS-PKK-LRx) (Ionis) - Profylaxe
  - Phase II resultaten gepubliceerd maart 2022
  - The study demonstrated a mean reduction of 97% in the number of monthly HAE attacks in weeks five to 17.
  - Hoge AE-QoL score
  - In weeks five to 17, 92% of patients treated with IONIS-PKK-LRx were attack-free

- STAR-0215 (Astria, eerst Catabasis)
  - NDA mid-2022, Phase I trial eind 2022, Phase 1b/2 results end 2023
  - Monoclonal antibody inhibitor of plasma kallikrein for HAE
  - Toediening eenmaal per drie maanden
  - QLS-215 has the potential to be the most patient-friendly chronic treatment option
  - New preclinical data for STAR-0215 in a presentation titled “Development of STAR-0215: An Engineered IgG1 Monoclonal Antibody Targeting Plasma Kallikrein for the Prevention of HAE” at the 2022 Fc Receptor and IgG Targeted Therapies Conference which takes place April 27th 2022 3.00pm in Boston, Massachusetts.

- BCX7353 (Biocryst) – Acuut
  - Orale liquid toediening. Fase III te starten

- ATN-249 (Attune) – Profylaxe
  - Orale toediening, kallikrein inhibitor. Phase I

- Garadacimab (CSL Behring) - Profylaxe - Recombinant monoclonal antibody
  - Subcutane toediening om de 4 weken
  - Garadacimab is a first-in-class, fully human, immunoglobulin G4 monoclonal antibody
  - Phase II afgerond
  - Mean percentage reductions were 88.68%, 98.94%, and 90.50% in three garadacimab groups - 75, 200, and 600 mg subcutaneous (SC) - versus placebo

- ALN-F12 (Alnylam) - Profylaxe
  - Subcutane toediening

- ARC-F12 (Arrowhead Pharmaceuticals) - Profylaxe
  - Subcutane toediening

- Verseon (Verseon) - Profylaxe
  - Orale toediening - Klein molecuul plasma kallikrein inhibitor

Gentherapieën

- CRISPR/Cas9 NTLA-2002 (Intellia Therapeutics) - Gentherapie
  - Eenmalige toediening
  - Plans to present interim data from ongoing first-in-human study of NTLA-2002 for hereditary angioedema (HAE) in2H 2022
  - Eerste patiënt behandeld
  - Juni 2021: Clinical Trial Application (CTA) to the New Zealand Medicines and Medical Devices Safety Authority for NTLA-2002 to initiate a first-in-human study
  - Meer autoriteiten aanvragen om studie uit te breiden naar meerdere landen
  - Aims to reduce plasma kallikrein activity to prevent excess bradykinin production leading to HAE attacks after a single course of treatment

- BMN-331 (Biomarin) - Gentherapie
  - Enrollment is open for the open-label, Phase 1/2 HAERMONY trial to evaluate the safety and effectiveness of BMN 331
  - Eenmalige toediening - IND-enabling studies in juli 2020 gestart
  - IND for BMN 331 was recently cleared by FDA and is active.
  [ul][li]ADVM-053 (Adverum) - Gentherapie
  Eenmalige toediening - FDA Orphan Drug Status

- RGX-314 (Regenxbio) - Gentherapie
  - Eenmalige toediening - samenwerking met Neurimmune bouwt voort op veelbelovende resultaten met ons klinische RGX-314-programma
  - A gene therapy product candidate utilizing NAV Vectors designed to deliver a gene encoding a therapeutic antibody targeting and binding to plasma kallikrein