Quarterly Shareholder Update – September 2017

Dear Shareholder,

This quarter saw several pleasing developments for Pharmaxis, not least of which the banking of a significant cash payment for one of our drug discoveries.

In August Pharmaxis received $27m from Boehringer in a milestone payment for the drug candidate we sold in 2015. This event was closely followed by confirmation that a phase 2 study into a second indication would be pursued by Boehringer before the end of 2017 that will trigger a further payment of $14m to Pharmaxis when it recruits its first patient.

The clinical programs are in diseases that are the result of diabetes complications; NASH and diabetic retinopathy, with Pharmaxis to continue to receive milestone payments from both programs as the studies progress. These two events are significant because they underscore the foresight of our drug discovery team to correctly identify that inhibiting the SSAO enzyme would provide such a valuable opportunity, and their ability to design a drug which Boehringer clearly feels has the properties suitable for development. This drug is now a key asset in Boehringer's strategy to target the complications of diabetes and the decision to pursue two different indications in parallel underlines their belief in this drug and their willingness to invest significant resources to maximize the potential benefits for patients.

Since the restructuring of the company in 2013 the Pharmaxis Drug Discovery team has been extraordinarily productive with the Boehringer partnered asset being followed up by our very promising anti-fibrotic LOXL2 drug candidates which are phase 1 ready and two earlier stage programs that we hope to progress to phase 1 next year. The Pharmaxis Board and management team have at the same time looked to further strengthen our capabilities in this critical part of the Pharmaxis business model.

Earlier this year we appointed Dr. Kathleen Metters to our Board. Kathleen was previously the Worldwide Head of Basic Research at Merck (MSD) and been CEO of a successful US Biotech company. The recently announced additional appointment of Kathleen to our Scientific Advisory Board along with the distinguished Australian academic and biotech entrepreneur Professor Darren Kelly, provides the Pharmaxis management team with unparalleled access and advice from leading academics and clinicians in our chosen fields of inflammation and fibrosis.

In a related move and after an extensive search we also appointed Dr. Dieter Hamprecht to the position of Head of Chemistry. Dieter has had a particularly distinguished career as a medicinal chemist, most recently with Boehringer Ingelheim in Milan. You can read more about him and the Scientific Advisory Board in this update.

I'm now very much looking forward to this next quarter as we return to the clinic with the LOXL2 program and begin the partnering process for that asset in earnest.

Sincerely,

Chief Executive Officer

Drug discovery

Boehringer Ingelheim initiates two phase 2 clinical trials for BI 1467335 (formerly known as PXS-4728A)

In August 2017 Boehringer Ingelheim commenced a 12 week, phase 2a proof of clinical principal trial in NASH of the drug candidate BI 1467335 acquired from Pharmaxis in 2015. The dosing of the first patient triggered a milestone payment to Pharmaxis of €18 million (A$27 million).

In September 2017 Boehringer Ingelheim announced it was initiating a phase 2a study in diabetic retinopathy (DR) of BI 1467335, marking the beginning of a clinical development program for a second indication– both studies targeting a severe diabetes complication.

Under the deal signed in 2015, Boehringer has total responsibility for the development program and Pharmaxis receives payments for multiple indications. Expanding the development plan to include diabetic retinopathy as well as NASH means that Pharmaxis will receive a €10m milestone payment when the first patient is dosed in the DR phase 2a study and that all the potential development milestones in the deal (€419m /A$625m), would be payable to Pharmaxis should both indications be approved. Developments in the quarter reaffirmed Pharmaxis choice of Boehringer as a partner that would seek to maximize the potential of the drug.

Both phase 2a clinical trials are expected to report in the second half of 2018.

LOXL2 inhibitor program set to commence phase 1 clinical trials

In September Pharmaxis and its collaborator UK biotechnology company Synairgen plc (AIM: SNG) announced completion of the preclinical development stage of their anti-fibrotic Lysyl Oxidase type 2 (LOXL2) inhibitor program allowing the program to commence human clinical phase I studies in Q4 2017.

The Pharmaxis drug discovery group has developed a number of selective small molecule inhibitors to the LOXL2 enzyme utilising the same amine oxidase platform that delivered BI-1467335. The LOXL2 enzyme is fundamental to the fibrotic cascade that follows chronic inflammation in the liver disease NASH, cardiac fibrosis, kidney fibrosis, and idiopathic pulmonary fibrosis (IPF), and it also plays a role in some cancers.

An extensive pre-clinical program performed on the program compounds confirmed they have all the characteristics of a successful once a day, oral drug, showing excellent efficacy in several different in vivo fibrosis models including fibrosis of the liver, lung, kidney and heart. In regulatory toxicity studies, the compounds have been well tolerated and shown a good safety profile.

Partnering plan for LOXL2 program

Pharmaxis has been discussing its LOXL2 program with large pharma companies since the beginning of 2016. There is interest in the program as it is one of the very few truly anti-fibrotic mechanisms in clinical development.

In parallel with the commencement of phase 1 clinical trials, Pharmaxis will enable scientific due diligence of the program by select large pharma companies interested in subsequent partnering discussions.

Drug development pipeline – other programs

The Pharmaxis amine oxidase chemistry platform has delivered five lead candidates in the last four years. In addition to the SSAO inhibitor acquired by Boehringer in 2015 and the two compounds developed in the LOXL2 program, the drug discovery team has advanced two other programs that are now in the final stages of pre-clinical testing and about to commence the toxicity studies that are prerequisite to phase 1 trials – expected to commence in 2018:

- a drug inhibiting both myeloperoxidase (MPO) and SSAO with potential anti-inflammatory application in respiratory and cardiovascular disease.
- a drug inhibiting all the LOX family of enzymes with potential anti-fibrotic application in scarring and severe fibrotic indications including some cancers.

Pharmaxis strengthens drug discovery capability

After a wide ranging recruitment program Pharmaxis has appointed Dr Dieter Hamprecht as the Head of Chemistry in the Pharmaxis Drug Discovery team, reporting to Department Head Dr Wolfgang Jarolimek. Dr Hamprecht was previously the Managing Director of Boehringer Ingelheim's Research group in Milan and has a distinguished career as a medicinal chemist having held senior positions at GSK and Boehringer with 37 publications and 44 patents to his name. He has a wide ranging experience in small molecules and peptides after 20 years working in multinational pharma company research groups and managing internal programs and assessing external opportunities as part of Boehringer's scientific diligence teams. His appointment reinforces the strength and depth of the Pharmaxis drug discovery team.

Pharmaxis also expanded its Scientific Advisory Board during the quarter with the appointment of Professor Darren Kelly and Dr Kathleen Metters.

Professor Kelly is the Associate Dean (Innovation and Enterprise, MDHS) at The University of Melbourne, the Director of Innovation and Enterprise at the Centre for Eye Research Australia (CERA) and Director of Biomedical Research in the Department of Medicine, St Vincent's Hospital Melbourne. Professor Kelly was the CEO of Fibrotech which was successfully sold to Shire in 2014 and is currently the CEO and Managing Director of Australian biotech company Occurx and the entrepreneur in residence at the Medical Research Commercialisation Fund. He has published over 200 manuscripts in the field of translational research and novel interventions many of which have had a direct impact on human disease.

Dr Metters was appointed as a non-executive director to the Pharmaxis Board in June 2017, and was subsequently appointed to the Scientific Advisory Board. Dr Metters has more than 25 years of experience in the discovery and development of novel therapies for the treatment of serious diseases. She spent 9 years with Merck & Co. including a period as senior vice president and head of Worldwide Basic Research and leading their External Discovery and Preclinical Sciences. She subsequently was appointed as President and Chief Executive Officer for Lycera Corp., a biopharmaceutical company pioneering innovative approaches to novel oral medicines for treatment of autoimmune diseases and cancer.

Further information concerning the Scientific Advisory Board is available on the Pharmaxis website.

Bronchitol and Aridol

Bronchitol® is an inhaled dry powder for the treatment of cystic fibrosis (CF) and has been the subject of three large scale global clinical trials conducted by Pharmaxis. The product is approved and marketed in Europe, Russia, Australia and several other countries.

Aridol® is an innovative lung function test designed to help doctors diagnose and manage asthma. Aridol is approved for sale in Australia, major European countries, the United States and South Korea.

United States

Subsequent to reporting positive top line results of the international multicentre clinical trial of Bronchitol (CF303) in June 2017, the clinical team has been focused on finalising the complete study report. This will shortly be delivered to the Company's US partner Chiesi who are responsible for completing and filing the New Drug Application with the FDA and the commercialisation of Bronchitol in the United States.

Subject to approval, Pharmaxis will receive a US$10 million milestone on the commercial launch of Bronchitol in the US, mid to high teen percentage royalties and will be the exclusive supplier of Bronchitol for the US market.

Western Europe

In the EU, Pharmaxis has appointed Chiesi as its exclusive Bronchitol distributor for the markets of the UK, Ireland and Germany and from May 2017 Chiesi became exclusive distributor for the Italian market. As expected Chiesi purchased Bronchitol for the UK and the Italian markets in the September quarter with orders for Germany and additional Italian inventory to ship in the December quarter.

Pharmaxis also markets Bronchitol in Austria, Denmark and Norway via its German based logistics provider, and Spain via an exclusive

distributor. Sales for Western Europe in the quarter were $457,000 compared to $180,000 in the September 2016 quarter, the increase reflecting increased sales to Chiesi for the UK and Italy.

Other territories

Bronchitol is sold in Australia by Pharmaxis and in Turkey and Russia by exclusive distributors.

Russia represents a potential significant opportunity for Bronchitol with approximately 4,000 CF patients on the Russian Cystic Fibrosis Registry and an annual market (2015) for CF drugs to deal with mucus clearance of approximately US$29 million. Following the receipt of approval in 2016 for both adult and paediatric CF patients, Pharmaxis has been navigating the process to have Bronchitol reimbursed nationally. Whilst the national reimbursement process is taking longer than initially anticipated, with support from CF clinics and constructive discussions with reimbursement authorities Pharmaxis has already started to receive approvals for individual patient reimbursement applications.

In August 2017 Pharmaxis received a positive recommendation from the Australian Pharmaceutical Benefits Advisory Committee (PBAC) for expanded reimbursement of Bronchitol for the treatment of cystic fibrosis. The submission for broader access to Bronchitol was strongly supported by the cystic fibrosis community. Pharmaxis has since reached in principle agreement with the Australian government to convert the PBAC recommendation into a listing on the Pharmaceutical Benefits Scheme in the near future.

Aridol

Aridol sales for the quarter were A$443,000 compared to A$505,000 in the September 2016 quarter, with the decrease primarily due to a lower level of sales to Korea in the current quarter.

Pharmaxis has appointed North American specialty pharmaceutical company Methapharm Inc as its exclusive distributor for the United States and is working towards reintroducing Aridol to the United States in 2018. Methapharm has been selected due to its extensive experience in the markets in which Aridol competes.

Pharmaxis is also working towards the launch of Aridol in Canada in 2018.

Corporate

2017 Annual General Meeting

The 2017 Annual General Meeting will be held on 13 November 2017 at the Christie Conference Centre, 3 Spring Street, Sydney, at 2.30 pm (Sydney time). The Notice of Meeting and Proxy Voting Form were distributed to shareholders on 12 October. The formal part of the Meeting will cover consideration of the Company's financial statements and remuneration report, the re-election of two non-executive directors and the grant of securities to the Chief Executive Officer.

CEO interviews

To view recent interviews with Pharmaxis CEO Gary Phillips discussing the Company's capability and approach to drug discovery visit the website at http://www.pharmaxis.com.au/investor-centre/new-investor-centre-page/

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Financials

Key financial metrics

A$'000	Three months ended	
(unaudited)	**30-Sept-17**	**30-Sept-16**
Income statement items		
Sale of Bronchitol & Aridol	1,049	897
Sale of drug candidate	26,891	-
Total revenue	**$28,911**	**$5,010**
Total expenses	(7,572)	(9,095)
Net profit (loss) after tax	**$21,347**	**($4,085)**
Segment results – adjusted EBITDA		
Bronchitol & Aridol	(1,460)	(1,452)
New drug development	24,498	(1,118)
Corporate	(981)	(1,228)
Total adjusted EBITDA	**$22,057**	**($3,798)**
Statement of cash flows		
Cash inflow/ (outflow) from:		
Operations	17,662	(6,173)
Investing activities	(110)	(150)
Financing activities	(427)	(426)
Total cash generated/(used)	**$17,125**	**($6,749)**
Cash at bank	**$38,629**	**$32,460**

Highlights

- The Company received $26.9 million during the quarter when the drug candidate sold to Boehringer Ingelheim in 2015 commenced a phase 2a clinical trial in NASH.
- Sales revenue for the quarter increased 17% over the comparable quarter in 2016.
- Net profit after tax for the quarter was $21.3 million compared to a loss of $4.1 million in the September quarter of 2016.
- Total expenses decreased by 17% with underlying core expenses for the quarter in the main unchanged from the comparable period. The following specific items accounted for the net decrease in total expenses of $1.5 million.
    - Clinical trial expenses in relation to clinical trial CF303 decreased by $3.0 million.
    - Drug development expenses increased by $1.0 million reflecting increased levels of research activity in several projects.
    - Other expenses include the net transfer of manufacturing labour and overhead to and from inventory as product is first manufactured and then subsequently sold to distributors and customers. Other expenses were a credit of $529,000 in the September 2016 quarter and $113,000 in the current quarter.

- Pharmaxis finished the quarter with $38.6 million. Net cash generated during the quarter was $17.1 million. Note that Pharmaxis has been funding clinical trial CF303 since Chiesi completed its contribution of US$22 million in December 2016 - cash used in the current quarter was A$2.8 million with approximately A$0.5 million remaining to be paid in the December 2017 quarter.
- Additional cash expected to be received in the current financial year includes:
  - The 2017 R&D tax credit of $3.1 million.
  - A milestone of $15 million from Boehringer Ingelheim when the first patient is dosed in a phase 2a clinical trial in diabetic retinopathy. The trial is currently recruiting subjects.

Segment information

A$'000	Segment information - three months ended							
(unaudited)	30-Sept-17				30-Sept-16			
Income statements	Bronchitol & Aridol	New drug developm't	Corporate	Total	Bronchitol & Aridol	New drug developm't	Corporate	Total
Revenue								
Sale of Bronchitol	605			605	392			392
Sale of Aridol	444			444	505			505
	1,049			**1,049**	**897**			**897**
Sale of drug candidate		26,891		26,891				
Clinical reimbursement	681			681	3,576			3,576
Tax credit	-	-	-	-	-	-		-
Other revenue	88	-	109	197	8	224	80	312
	1,818	**26,891**	**109**	**28,818**	**4,481**	**224**	**80**	**4,785**
Expenses								
Employee costs	(1,428)	(577)	(508)	(2,513)	(1,485)	(508)	(647)	(2,640)
Clinical trials	(776)	(199)		(975)	(3,770)	-	-	(3,770)
Drug discovery	-	(1,544)		(1,544)	-	(732)	-	(732)
Other expenses	(1,074)	(73)	(582)	(1,729)	(678)	(102)	(661)	(1,441)
Total expenses	(3,278)	(2,393)	(1,090)	(6,761)	(5,933)	(1,342)	(1,308)	(8,583)
Adjusted EBITDA	**($1,460)**	**$24,498**	**($981)**	**$22,057**	**($1,452)**	**($1,118)**	**($1,228)**	**($3,798)**

- Sale of Bronchitol increased due to increased sales to Chiesi for the UK and the first shipment in relation to the Italian market.
- Sale of Aridol reduced primarily because of a lower level of sales to Korea in the current quarter.
- Clinical trial reimbursements and clinical trial costs reduced following reporting of study CF303 in June 2017. A further $500,000 of reimbursement revenue and clinical trial costs will be booked in the December 2017 quarter.
- Increased new drug development expenses for the quarter reflect additional expenditure on the LOXL2 program as it approached phase 1 studies ($0.9 million for the quarter) and additional expenditure on other pipeline assets including the LOX program ($0.6 million for the quarter).
- Bronchitol & Aridol Other Expenses increased mainly due to the change in net transfer of labour and overhead into inventory as discussed above.

Income statements

A$'000	Three months ended	
(unaudited)	**30-Sept-17**	**30-Sept-16**
Revenue		
Sale of Bronchitol & Aridol	1,049	897
Sale of drug candidate	26,891	-
Clinical trial cost reimbursements	681	3,577
Interest	93	224
Drug discovery service fee	-	224
Other	197	88
Total revenue	**$28,911**	**$5,010**
Expenses		
Employee costs	(2,817)	(2,892)
Administration & corporate	(592)	(536)
Rent, occupancy & utilities	(277)	(244)
Clinical trials	(975)	(3,770)
Drug development	(1,544)	(732)
Sales, marketing & distribution	(246)	(207)
Safety, medical and regulatory affairs	(184)	(375)
Manufacturing purchases	(465)	(314)
Other	(25)	311
Depreciation & amortisation	(781)	(758)
Foreign currency exchange gains & losses	482	584
Finance expenses	(148)	(162)
Total expenses	**(7,572)**	**(9,095)**
Net profit (loss) before tax	**21,339**	**(4,085)**
Income tax (expense)/credit	8	-
Net profit (loss) after tax	**$21,347**	**($4,085)**

Summary balance sheets

A$'000		
(unaudited)	**30-Sept-17**	**30-June-17**
Assets		
Cash	38,629	21,504
R&D tax credit receivable	3,100	3,100
Accounts receivable	1,520	1,262
PP&E	14,143	14,860
Other	4,913	4,708
	$62,305	**$45,434**
Liabilities		
Accounts payable and accrued expenses	2,746	6,134
Lease liability (Frenchs Forest facility)	9,017	9,251
Financing agreement (not repayable other than as a % of US & EU Bronchitol revenue)	21,675	22,141
Other liabilities	3,696	4,387
	$37,134	**$41,913**