Quarterly Shareholder Update – June 2019

Dear Shareholder,

Sustainability is now an established goal in the way we live whether it be by the recycling of waste, water consumption or a move to cleaner energy. Sustainability, however, is not always considered in relation to biotech investments where many companies have one asset under development that will lead inevitably to either a profitable exit or a complete loss. Pharmaxis is different in that we champion a portfolio approach to building value and mitigating risk. The June quarter saw many of our portfolio assets move forward, adding value that can be realized down the track at multiple time points, rather than one "Russian roulette" moment.

The June quarter saw one of our long-term pipeline assets reach a pivotal moment when the FDA Pulmonary and Allergy Drug Advisory Committee (PADAC) met to consider questions put to them by the FDA on the efficacy and safety of Bronchitol for the treatment of cystic fibrosis in adult patients. The positive vote led to a complete response letter from the FDA that provides a clear path to approval subject to the completion of a human factor study by Chiesi. You can read the details of that later in this newsletter but suffice to say we are confident that the FDA will have the information it requires to be able to approve Bronchitol by Q1 2020. The opening of the US market for Bronchitol builds on the successful relaunch of Aridol earlier this year. A US$10m Bronchitol milestone payment anticipated in Q2 2020 will be welcome and it will also be very gratifying to see the Bronchitol and Aridol franchise turn into a cash flow positive and sustainable business next year.

Another plank of sustainability for us is the potential cash flow from the deal struck with Boehringer for our first in class AOC3 inhibitor for the liver disease NASH. Boehringer have been very active acquiring a number of other NASH and anti-fibrotic assets in the last few months and several of you have asked my opinion on the ramifications for Pharmaxis. I'm actually pleased to see Boehringer's commitment to these disease areas. Most commentators and clinicians continue to believe that combination therapy will be needed to make significant inroads and indeed our deal with Boehringer anticipates and protects our interests should our drug be commercialized as part of a combination approach. Boehringer announced that it expects to report the results from the NASH phase 2a study in quarter 4 this year after completing both the study and internal post study analysis. Obviously a positive decision at that point opens the door to a phase 2b study and the next set of milestone payments payable at the start of phase 3.

I wrote in the March quarterly update that we had entered the commercial stage of partnering discussions for our LOXL2 inhibitor program and we have also been very active at various partnering forums including the BIO 2019 conference in Philadelphia in June. It is a complex and multi-faceted process, with the fact that there are a number of therapeutic indications under consideration and changing market conditions tending to extend the discussions. Confidentiality is fundamental to securing the right outcome for our shareholders so we will not provide any updates until we reach a conclusion to the process we have embarked on which I expect before year end.

Continuing with the sustainability theme, we have long practiced getting multiple opportunities to the point where they have a chance to demonstrate their value to patients and of course shareholders. It's important to remember this early stage research doesn't carry the heavy investments associated with later stage clinical studies and the support from the Australian Government in the form of an R&D tax incentive further improves its cost effectiveness. The $6m tax credit we expect to receive for FY2019 is allied to

unprecedented productivity of our drug discovery team who have managed to get four drugs of their own invention into the clinic in the last five years with one more on the way in 2020, representing a great return on investment.

We are storing up a great deal of value potential in our pipeline and it was gratifying to hear the enthusiasm for our clinical trial plans in myelofibrosis, pancreatic cancer and scarring from recent meetings with our Scientific Advisory Board and clinical key opinion leaders at the ASCO conference in Chicago. Our systemic pan LOX inhibitor for myelofibrosis and pancreatic cancer completed the first part of its phase 1 study in healthy volunteers and will continue with the second phase later this year with a view to commencement of the first study in patients first half next year.

Sincerely,

Gary Phillips – Chief Executive Officer

Drug discovery

Boehringer Ingelheim development of BI 1467335 (formerly known as PXS-4728A)

Boehringer Ingelheim is developing BI 1467335, a drug it acquired from Pharmaxis in 2015, for two indications – the liver disease Non-Alcoholic Steatohepatitis (NASH) and the eye disease diabetic retinopathy (DR). Boehringer initiated phase 2a proof of clinical principle trials for NASH in August 2017 and for DR in January 2018. The achievement of these development milestones resulted in Pharmaxis receiving a total of €28 million (A$42 million) in the 2018 financial year.

In June Boehringer advised that its phase 2(a) proof of clinical principle trials for NASH had been completed with the last patient dosed. Boehringer expects to report the results from this study to Pharmaxis in quarter 4 this year after it has completed an internal post study analysis as well as an internal review of the future NASH development program for BI 1467335.

Non-alcoholic fatty liver disease (NAFLD), the most common liver disorder in Western industrialised nations, and its more serious form NASH, is highly prevalent amongst patients with type 2 Diabetes. NASH is a major cause of liver fibrosis and cirrhosis and is an area of high unmet medical need with no treatments currently available. The high prevalence of type 2 diabetes and obesity is expected to make NASH one of the most common causes of advanced liver disorders in coming decades. 25% of the general adult population in the world has NAFLD and the prevalence of NASH has been found to range from 1.5% to 6.5% in current research, a number twice as high as 20 years ago.

The phase 2a NASH trial is a multi-centre, double-blind design in 114 patients with clinical evidence of NASH. The trial is being conducted in nine countries across North America and Europe. The primary objectives are to establish proof of clinical principle, investigate suitable dosing, and to evaluate the safety of BI 1467335. Patients have been randomised to either one of four dosages of BI 1467335 or to placebo for a 12-week treatment period followed by a 4-week observation period. A subsequent Phase 2b study will seek to confirm and extend these findings.

Diabetic retinopathy is the leading cause of vision-loss in adults. Of an estimated 285 million people with diabetes mellitus worldwide, approximately one third have signs of DR and of these, a further one third is vision-threatening. The DR trial is scheduled to complete early in the first half of 2020.

Boehringer has total responsibility for the development program of BI 1467335 and Pharmaxis receives payments upon achievement of certain development milestones. The total development milestones in the deal (€419m /A$625m), would be payable to Pharmaxis should both indications be approved.

LOXL2 inhibitor program

The Lysyl Oxidase Like 2 (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.

The Pharmaxis drug discovery group developed two small molecule inhibitors to the LOXL2 enzyme which have now completed phase 1 clinical trials and 3-month toxicology studies. In January 2019 Pharmaxis announced that for both compounds, doses that resulted in 85% or greater inhibition of the target enzyme in the phase 1 studies were below the human equivalent No Observed Adverse Effect Level doses in all toxicity studies and therefore demonstrated an adequate safety margin to start phase 2 studies of up to 3 months in length.

The Pharmaxis LOXL2 program is one of the very few truly anti-fibrotic mechanisms in clinical development. The excellent pharmacokinetic parameters and the significant and long lasting inhibition of the target LOXL2 enzyme showed that these compounds are best-in-class. Subsequent to completion of the data package including the phase 1 trials and the three month tox studies, the company has entered the commercial stage of partnering discussions with a number of pharmaceutical companies. Pharmaxis will provide more information when the process concludes.

Systemic LOX inhibitor program

In addition to the SSAO inhibitor (BI 1467335) and the LOXL2 program, Pharmaxis is progressing two lysyl oxidase (LOX) programs from its amine

oxidase chemistry platform, both of which are planned to partner after phase 2 clinical trials.

The most advanced LOX program has developed an oral once-a-day drug that inhibits all lysyl oxidase family members (LOX, LOXL1, 2, 3 & 4).

The compound successfully cleared pre-clinical safety and toxicity studies in the third quarter of 2018 and has shown significant reductions in fibrosis in in-vivo models of kidney fibrosis, lung fibrosis, myelofibrosis and pancreatic cancer. It is suited to the treatment of severe fibrosis as well as cancer with prominent stroma (connective tissue) or fibrotic metastatic niches.

In February 2019 Pharmaxis announced commencement of a Phase 1 clinical trial, consisting of two stages. The first single ascending dose stage (SAD) conducted in 40 healthy subjects completed in June 2019. The compound demonstrated a good pharmacokinetic profile, full engagement of the target enzymes and a good safety profile. The second multiple ascending dose stage (MAD) to be conducted in 16 healthy subjects will commence in the third quarter and report later in the year.

During this quarter the compound completed its three month tox studies and started its 6 months evaluation.

The Company aims to be ready to commence phase 1c/2 study in myelofibrosis and /or pancreatic cancer patients by first half 2020.

Topical LOX inhibitor program

The Company's other LOX program has developed a drug for topical application with the potential for use in scar revision, keloid scarring and scarring from burn wounds.

A lead candidate has been selected and is currently in pre-clinical development including initial stability of the topical formulation, ongoing evaluation in various disease models of scarring and tox studies.

The program aims to commence phase 1 studies in early 2020 and is planning to conduct the trial in healthy volunteers with scarring so as to be able to simultaneously test clinical efficacy.

Mannitol business

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.

There has been a lot of activity relating to both Bronchitol and Aridol during the quarter.

United States

The Company's US partner Chiesi Group (Chiesi) is responsible for the commercialisation of Bronchitol in the United States. Chiesi filed a resubmission of the Bronchitol New Drug Application (NDA) to the US Food and Drug Administration (FDA) in December 2018. Following a positive recommendation from a Pulmonary-Allergy Drugs Advisory Committee meeting convened by the FDA on 8 May 2019, Chiesi received a complete response letter from the FDA on 19 June 2019 detailing the remaining matters to be addressed before Bronchitol can be approved for adult cystic fibrosis patients in the United States. Based upon the clear and achievable path to approval communicated in the complete response letter, Pharmaxis believes that the FDA review of the Bronchitol NDA will be completed in Q1 2020.

The main requirement included in the FDA complete response letter is that Chiesi revise the product packaging and user instructions; and then conduct a human factor study demonstrating that the revised user components enable healthcare professionals to properly administer the mannitol tolerance test. These remaining requirements are targeted for completion by the end of 2019. Based on experience with healthcare professionals in other markets where the Company's training and packaging has supported thousands of mannitol tolerance tests that have been conducted to ensure patients hypersensitive

to mannitol are not prescribed Bronchitol, Pharmaxis is very confident the requested FDA changes can be efficiently implemented and will be effective in achieving the desired goal. Pharmaxis has been sharing its experiences in other markets with Chiesi and continues to work collaboratively to prepare for a successful introduction to patients in US cystic fibrosis clinics.

Subject to approval, Pharmaxis will receive a US$10 million milestone payment 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, Chiesi is the Pharmaxis exclusive Bronchitol distributor for the markets of the UK, Ireland, Germany and Italy. The Chiesi territory was recently expanded to include Norway, Sweden, Finland, Denmark, Cyprus and Greece.

During the quarter Pharmaxis appointed EffRx Pharmaceuticals SA, a commercial-stage company that commercialises niche and orphan medicines in Switzerland and Europe as its exclusive licensee for the registration and commercialisation of Bronchitol for cystic fibrosis in Switzerland. Under the terms of the agreement, EffRx will take responsibility for registering, obtaining pricing and reimbursement as well as commercialising Bronchitol in Switzerland.

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

Other territories

Bronchitol is sold in Australia by Pharmaxis and in Turkey, the Czech Republic and Russia by exclusive distributors.

Russia represents a potential significant opportunity for Bronchitol which was approved for both adult and paediatric cystic fibrosis patients in 2016. Subsequent to Bronchitol being reimbursed nationally from 1 January 2019, an order for 2,000 Bronchitol packs ($590,000) was sold to the Company's Russian distributor in June and in- market sales have been encouraging.

Bronchitol sales

Bronchitol sales for the three and twelve months ended 30 June 2019 and 30 June 2018 are as follows:

$'000	Three months		Twelve months	
	2019	2018	2019	2018
Australia	277	294	1,059	949
Western Europe	24	893	1,041	2,900
Russia & Eastern Europe	306	104	464	235
Total	**$607**	**$1,291**	**$2,564**	**$4,084**

There were no large shipments to the Company's European distributor during the quarter, with the next major order shipped in July.

Sales to the Company's Russian distributor in the quarter were reduced by a credit note of $309,000 issued with respect to expired product that was originally supplied in 2017 in anticipation of sales prior to receiving re-imbursement in January 2019.

Pharmaxis distributors typically order Bronchitol on a six monthly basis.

Aridol

Aridol was relaunched in the US in December 2018 by Pharmaxis' exclusive distributer in North America, Methapharm Inc., who are experts in the specialist respiratory diagnostic market. A further order was shipped to Methapharm in the current quarter.

In June 2019 Pharmaxis received approval for Aridol from Canadian regulatory authorities. The Company expects the product launch by Methapharm to occur in the second half of 2019.

Aridol sales

Aridol sales for the three and twelve months ended 30 June 2019 and 30 June 2018 are as follows:

$'000	Three months		Twelve months	
	2019	2018	2019	2018
Australia	131	121	471	430
Europe	246	246	979	902
USA	344		1,003	
South Korea	177	242	659	678
Total	**$898**	**$609**	**$3,112**	**$2,010**

Corporate

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Financials

Key financial metrics

A$'000	Three months ended		Twelve months ended	
(unaudited)	**30-Jun-19**	**30-Jun-18**	**30-Jun-19**	**30-Jun-18**
Income statements				
Sales of Bronchitol & Aridol	1,505	1,900	5,676	6,094
Milestones from sale of drug				42,130
Total revenue	7,797	2,213	13,080	50,833
Total expenses	(8,939)	(9,859)	(33,138)	(44,413)
Net profit (loss) after tax	**(1,141)**	**(7,646)**	**(20,058)**	**6,428**
Segment results – adjusted EBITDA				
Bronchitol & Aridol	(1,819)	(1,198)	(5,013)	(3,786)
New drug development	719	(3,682)	(6,764)	28,771
Corporate	(829)	(947)	(3,874)	(13,466)
Total	**(1,929)**	**(5,827)**	**(15,651)**	**11,519**
Statement of cash flows				
Cash inflow/ (outflow) from:				
Operations	(3,404)	(2,712)	(19,798)	12,206
Investing activities	(124)	(280)	(981)	(884)
Financing activities	(477)	(443)	20,830	(1,753)
Total cash generated/(used)	**(4,005)**	**(3,435)**	**51**	**9,569**
Cash at bank	**31,124**	**31,073**	**31,124**	**31,073**

Highlights

- Revenue
  - See above for detail and commentary on Bronchitol and Aridol sales.
  - The Company has booked an R&D tax incentive of $6.0m in respect of the 2019 financial year. Payment of the credit is expected in the second half of 2019.
  - In comparing the revenue with the prior comparable periods please note the milestone from sale of drug in the March 2018 quarter related to a milestone received from Boehringer Ingelheim on the commencement of a phase 2a clinical trial in diabetic retinopathy.
- Expenses
  - The reduction in total expenses compared to the prior quarter is primarily due to lower unrealised foreign currency exchange losses in the current quarter.
  - Total expenses for the comparable twelve months included $9.6 million of costs incurred in the December quarter of 2017 associated with changes to the collaboration agreement with Synairgen. Expenditure on clinical trials and drug development were approximately $1.8 million lower in the 2019 financial year.

- Cash
  - The Company finished the year with $31 million in cash, with the R&D tax incentive expected to be received later in CY 2019.

Segment information

A$'000	Segment information - three months ended							
(unaudited)	30-Jun-19				30-Jun-18			
Income statements	Bronchitol & Aridol	New drug development	Corporate	Total	Bronchitol & Aridol	New drug development	Corporate	Total
Revenue								
Sale of Bronchitol	607			607	1,291			1,291
Sale of Aridol	898			898	609			609
	1,505			**1,505**	**1,900**			**1,900**
Milestones from sale of drug								0
Tax credit		5,962		5,962		1		1
Other revenue	7		128	135	5		122	126
	1,512	**5,962**	**128**	**7,602**	**1,905**	**1**	**122**	**2,028**
Expenses								
Employee costs	(1,636)	(671)	(433)	(2,740)	(1,490)	(722)	(442)	(2,654)
Clinical trials		(1,118)		(1,118)	30	(1,175)		(1,145)
Drug discovery		(1,516)		(1,516)		(1,589)		(1,589)
Other expenses	(1,695)	(195)	(524)	(2,414)	(1,643)	(197)	(627)	(2,467)
Total expenses	(3,331)	(3,500)	(957)	(7,788)	(3,103)	(3,683)	(1,069)	(7,855)
Adjusted EBITDA	**($1,819)**	**$2,462**	**($829)**	**($186)**	**($1,198)**	**($3,682)**	**($947)**	**($5,827)**

Commentary for the quarter

- Bronchitol & Aridol:
  - Sales of Bronchitol and Aridol are discussed in commentary above.
  - Expenses for the quarter were consistent with the prior period.
- New drug development:
  - The Company has booked an R&D tax credit of $6.0 million in respect of the 2019 financial year. Payment of the credit is expected in the second half of 2019.
  - Clinical trial expenses relate to the phase 1 trial for the LOX oral program that commenced in the March quarter of 2019 ($1.1 million). In 2018 the clinical trial expenses related to the phase 1 trials conducted in the LOXL2 program which completed in the December 2018 quarter ($382,000).
  - Drug discovery expenses include work on the LOX oral program ($411,000 for the quarter; $296,000 in 2018) and the LOX topical program ($512,000 for the quarter; $106,000 in 2018).

A$'000	Segment information - twelve months ended							
(unaudited)	30-Jun-19				30-Jun-18			
Income statements	Bronchitol & Aridol	New drug development	Corporate	Total	Bronchitol & Aridol	New drug development	Corporate	Total
Revenue								
Sale of Bronchitol	2,564			2,564	4,084			4,084
Sale of Aridol	3,112			3,112	2,010			2,010
	5,676			**5,676**	**6,094**			**6,094**
Milestones from sale of drug						42,130		42,130
Clinical reimbursement					1,188			1,188
Tax credit		5,962		5,962		161		161
Other revenue	27		506	533	186	5	471	662
	5,703	**5,962**	**506**	**12,171**	**7,468**	**42,296**	**471**	**50,235**
Expenses								
Employee costs	(6,083)	(2,837)	(1,932)	(10,852)	(5,695)	(2,753)	(1,883)	(10,331)
Clinical trials	621	(2,975)		(2,354)	(160)	(3,465)		(3,625)
Drug discovery		(6,308)		(6,308)		(6,816)		(6,816)
Other expenses	(5,254)	(606)	(2,448)	(8,308)	(5,399)	(491)	(2,474)	(8,364)
Change in collaboration							(9,580)	(9,580)
Total expenses	(10,716)	(12,726)	(4,380)	(27,822)	(11,254)	(13,525)	(13,937)	(38,716)
Adjusted EBITDA	**($5,013)**	**($6,764)**	**($3,874)**	**($15,651)**	**($3,786)**	**$28,771**	**($13,466)**	**$11,519**

Commentary for the twelve months

- Bronchitol & Aridol:
  - Sales of Bronchitol and Aridol are discussed in commentary above. Note excluding the one-off credit note of $411,000 issued to the Company's Russian distributor, total sales for 2019 would have been the same as in 2018.
  - Clinical trial reimbursements and clinical trial costs ceased following completion of study CF303 in 2017.
  - Positive clinical trials expense consisted of a $621,000 unexpected refund from the clinical research organization that managed the CF303 clinical trial.
- New drug development:
  - The milestones from sale of drug in the prior period relate to two milestones received from Boehringer Ingelheim on the commencement of phase 2a clinical trial in NASH and diabetic retinopathy.
  - The Company has booked an R&D tax credit of $6.0 million in respect of the 2019 financial year.
  - Clinical trial expenses relate to the phase 1 trials being conducted in the LOXL2 program which completed in the December 2018 quarter ($1.4 million; $3.4 million in 2018)) and the phase 1 trial for the LOX oral program that commenced in the March quarter of 2019 ($1.6 million).
  - Drug discovery expenditure for the period includes the LOXL2 program ($1.0 million compared to $2.3 million in 2018), the LOX oral program ($2.2 million compared to $1.8 million in 2018) and the LOX topical program ($1.5 million compared to $100,000 in 2018).
- Corporate:
  - Note the $9.6 million of costs incurred in 2018 associated with changes to the collaboration agreement with Synairgen.

Income statements

A$'000	Three months ended		Twelve months ended	
(unaudited)	**30-Jun-19**	**30-Jun-18**	**30-Jun-19**	**30-Jun-18**
Revenue				
Revenue from sale of goods	1,505	1,900	5,676	6,094
Milestones from sale of drug				42,130
Clinical trial cost reimbursements				1,188
Interest	195	185	909	598
Drug discovery service fee				
R&D tax incentive	5,962	1	5,962	161
Other	135	127	533	662
Total revenue	**$7,797**	**$2,213**	**$13,080**	**$50,833**
Expenses				
Employee costs	(2,854)	(2,949)	(11,928)	(11,531)
Administration & corporate	(492)	(488)	(2,179)	(2,310)
Rent, occupancy & utilities	(348)	(349)	(1,386)	(1,279)
Clinical trials	(1,119)	(1,145)	(2,354)	(3,625)
Drug development	(1,516)	(1,589)	(6,308)	(6,816)
Sales, marketing & distribution	(375)	(266)	(1,136)	(1,163)
Safety, medical and regulatory affairs	(280)	(312)	(896)	(885)
Manufacturing purchases	(443)	(533)	(1,374)	(1,774)
Other	(509)	(518)	(1,380)	(1,134)
Depreciation & amortisation	(665)	(760)	(2,619)	(3,112)
Foreign currency exchange gains & losses	(225)	(818)	(1,340)	(641)
Finance costs	(113)	(132)	(238)	(563)
Costs in relation to change in collaboration agreement				(9,580)
Total expenses	**(8,939)**	**(9,859)**	**(33,138)**	**(44,413)**
Net profit (loss) before tax	**(1,142)**	**(7,646)**	**(20,058)**	**6,420**
Income tax credit/(expense)				8
Net profit (loss) after tax	**($1,142)**	**($7,646)**	**($20,058)**	**$6,428**

Summary balance sheets

A$'000		
(unaudited)	**30-Jun-19**	**30-June-18**
Assets		
Cash	31,124	31,073
R&D tax credit receivable	5,962	-
Accounts receivable	1,171	1,787
Inventory	2,116	2,398
PP&E	10,262	12,451
Other	2,033	2,388
	$52,668	**$50,097**
Liabilities		
Accounts payable and accrued expenses	4,194	4,926
Lease liability (Frenchs Forest facility)	7,171	8,268
Financing agreement (not repayable other than as a % of US & EU Bronchitol revenue)	23,626	22,754
Other liabilities	2,863	3,031
	$37,854	**$38,979**
Net Assets	**$14,814**	**$11,118**