Quarterly Shareholder Update – June 2018

Dear Shareholder,

There are times in the life of a research based company when progress can be slow; waiting for late phase clinical studies to recruit patients is but one example. However, this last quarter at Pharmaxis has been the exact opposite. We have data from the phase 1 studies in our LOXL2 program coming at us daily, ongoing discussions with multiple interested companies in that program further energised at the June BIO partnering conference, and turning points in some programs within our pre-clinical pipeline that suggest great things to come. You will find details on all the above in this quarterly update but let me briefly cover the high points:

The Pharmaxis Drug Discovery team working within our amine oxidase chemical platform identified two lead candidates that inhibit the fibrotic enzyme LOXL2. After extensive pre-clinical testing these compounds were deemed fit to go into human clinical development. The great thing about phase 1 studies is that cohorts of healthy subjects are small, dosing is short, and we see a lot of the data from these groups as it is generated. We announced the initial findings from the single ascending dose (SAD) stage during the quarter. This is still early days but both drugs so far have good safety profiles, behave in a predictable fashion as the dose is increased and most significantly we have managed to measure their impact on the target LOXL2 enzyme in human serum. Both compounds achieved a significant and long lasting inhibition which positions them as best in class drugs, differentiating them from the competition of other small molecule drugs and antibodies which have failed to demonstrate this same essential feature.

Good inhibition of the enzymatic activity for 24 hours after a single dose made the discussions held with potential partners at BIO 2018 in Boston last month much more interesting. We have seen an increase in the number of companies wanting to know more about the Pharmaxis program and also an increase in the intensity of those meetings as the profile of our drugs becomes apparent. There have been numerous publications validating the positive correlation between the levels of LOXL2 enzyme and the advancement of fibrosis in a number of diseases such as IPF and NASH. There is a strong case to study LOXL2 inhibition in later stage clinical trials and, for those companies supporting that view, we believe we have the best drug candidates. The data from the multiple ascending dose stage of these trials will continue to come in during the next two quarters but we can already see from the initial results that the excellent profiles seen in the SAD stage appear to be maintained. Partnering these drugs in a global deal with a large Pharma company is the number one objective for the Pharmaxis team in the second half of the year.

It's very rewarding to see programs mature and reach a point where their value can be realised. It's also exciting to see new ideas and concepts suddenly start to gain real traction and emerge as the potential next generation of pipeline drugs become ready for the clinic. I will have more to say on this topic in the next quarterly update, but the early stage proof of concept studies and initial safety data suggest that there are two more programs from the Pharmaxis amine oxidase platform that will generate drugs to put into the clinic next year. We are still evaluating which particular diseases might best suit these new drugs, but the shortlist contains some which have no current effective treatments and these drug candidates have already attracted interest from key opinion leading clinicians in areas such as pancreatic cancer where discussions to commence a phase 1 study in patients are already taking place.

Sincerely,

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 – NASH and diabetic retinopathy (DR). Boehringer initiated phase 2a proof of clinical principle trials for both development programs in 2017. Pharmaxis received €18 million (A$27 million) when the NASH trial dosed its first patient in August 2017 and €10 million (A$15 million) when the DR trial dosed its first patient in January 2018.

Both trials are due to complete in the first half of 2019.

NASH is an area of significant interest to large pharma companies and in addition to BI 1467334, Pharmaxis has a LOXL2 inhibitor under development for NASH, as outlined below.

DR 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 of DR is vision-threatening.

Under the deal signed in 2015, Boehringer has total responsibility for the development program and Pharmaxis receives payments for multiple indications. The total development milestones in the deal (€419m /A$625m), would be payable to Pharmaxis should both indications be approved.

We eagerly await the results of these first trials of efficacy in humans.

LOXL2 inhibitors in the clinic

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.

The Pharmaxis drug discovery group developed two small molecule inhibitors to the LOXL2 enzyme which were well tolerated in regulatory toxicity studies with a good safety profile.

The two compounds have both cleared the first stage of phase 1 studies where single oral doses of different strengths were trialed in healthy volunteers. Key results included:

- Both compounds were well tolerated and no safety signals were detected
- Pharmacokinetic (PK) parameters of both compounds increased with ascending dose
- The proprietary target engagement assay indicated that both compounds inhibited the LOXL2 enzyme in a dose-related fashion
- 24 hour inhibition of the LOXL2 enzyme was achieved with a single dose

Both drugs have now progressed to the final stage where different fixed doses are given for 14 days and dosing for one compound has been completed. Both studies will complete this quarter. Preliminary data confirms the results of the first stage with no additional safety concerns seen to date and the dose dependent PK profile maintained. Target engagement data is limited at this time but appears to mimic the positive findings from the first stage data very closely.

Pharmaxis has also initiated 3-month tox studies to further de-risk the program and add more value for prospective pharmaceutical company partners.

Pharmaxis has conducted an extensive pre-clinical program on the compounds. As research into predictive in vivo animal models for anti-fibrotic diseases such as NASH and IPF and their biomarkers continue, the compounds show excellent efficacy in several different in vivo fibrosis models including fibrosis of the liver, lung, kidney and heart.

Partnering plan for LOXL2 program

Large pharma is interested in the Pharmaxis LOXL2 program as it is one of the very few truly anti-fibrotic mechanisms in clinical development and a number of pharma companies have been following the progress of the Pharmaxis LOXL2 program for more than two years. At the recent BIO18 partnering conference in Boston the Company briefed potential partners on data emerging from the phase 1 clinical studies and the formal due diligence process.

Pharmaxis plans to partner the LOXL2 program in the second half of 2018 after the phase 1 trials and longer-term toxicity studies report.

Drug development pipeline – other programs

In addition to the SSAO inhibitor (BI 1467335) and the LOXL2 program, Pharmaxis has other programs developed from its amine oxidase chemistry platform. Lead candidates have been identified in two of these programs and both have commenced the toxicity studies that are prerequisite to commencing phase 1 clinical trials with the objective of being phase 1 ready for one of the programs within the next six months.

- An oral drug inhibiting all lysyl oxidase family members with potential anti-fibrotic application in severe fibrotic indications. This candidate has positive results in in vivo models of myelofibrosis and pancreatic cancer and additional confirmatory studies are being run at the present time.
- a dual acting drug inhibiting SSAO and myeloperoxidase (MPO) for the treatment of inflammation.

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

The Company's US partner Chiesi is responsible for the commercialisation of Bronchitol in the United States and is currently preparing to resubmit a New Drug Application with the FDA. Chiesi expect to complete and resubmit the NDA in the December quarter of 2018.

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.

Pharmaxis also markets Bronchitol in Austria, Denmark and Sweden via its German based logistics provider, and Spain via an exclusive distributor. Sales for Western Europe in the quarter were $893,000 (compared to $35,000 in the June 2017 quarter) and $2.9 million for the year (compared to $1.1 million in 2017).

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. 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 and a recent submission is expected to be reviewed in the second half of calendar 2018. The Company has been successful in obtaining approval for several hundred patients to receive Bronchitol via an individual reimbursement scheme. The Company's Russian distributor is supplying Bronchitol to a number of these patients and has orders from clinics for further supply. There were no sales by the Company to its Russian distributor during the quarter.

In the Czech Republic, Bronchitol has secured marketing and reimbursement approval and the first sale to the Company's distributor was shipped during the current quarter ($129,000).

Sales in Australia of $294,000 for the quarter compared to $170,000 in the June 2017 quarter. This is the second quarter of increased sales and follows the widened government reimbursement for Bronchitol granted on Jan 1st 2018.

Aridol

Aridol sales for the quarter were A$609,000 compared to A$589,000 in the June 2017 quarter, following increased sales in Australia. Together with its US and Canadian Aridol distributor Methapharm Inc, Pharmaxis has filed a regulatory submission with the US FDA to enable Aridol to resume sales in the USA. The US FDA is expected to respond later this year.

A submission to Canadian authorities was filed this quarter and the approval process is expected to take approximately 12 months.

Corporate

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Interviews with Pharmaxis director Dr Kathleen Metters

Dr Kathleen Metters is a global pharmaceutical industry executive with more than 25 years' experience and, since June 2017, has served on the Pharmaxis Board. Dr Metters visited Sydney in May 2018 for a Pharmaxis Scientific Advisory Board meeting and the annual Pharmaxis Board strategic review.

In an interview with Proactive Investor Dr Metters said she has been extremely impressed with the science and business focus at Pharmaxis noting, "This is an incredibly innovative research group. They have moved into pre-clinical development compounds for particular therapeutic targets to test new mechanisms that have never been done in the industry before. They have all the qualities to do it again and again. They are really rigorous scientists, everything they do is of the highest quality. They know every step of the business. This is a small company but it goes from research to commercialisation so they have really exquisite line of sight on what they need to do to bring forward new therapies. They are intensively collaborative…and all this adds up to the "secret sauce" which is the perfect culture.

Watch the interview here.

In its 22 July issue, industry news publication Bioshares features an interview Dr Kathleen Metters who notes the Company has "a very deep understanding of what makes a good molecule. That's just years of experience. I've been impressed by the way that Pharmaxis has built outstanding relationships with KOLs and outside experts."

Read the interview on the Pharmaxis website.

Financials

Key financial metrics

A$'000	Three months ended		Twelve months ended	
(unaudited)	**30-June-18**	**30-June-17**	**30-June-18**	**30-June-17**
Income statements				
Sales of Bronchitol & Aridol	1,900	866	6,094	4,823
Milestones from sale of drug	-	-	42,130	-
Total revenue	2,213	6,945	50,833	18,001
Total expenses	(9,857)	(11,057)	(44,413)	(36,347)
Net profit (loss) after tax	**(7,645)**	**(4,112)**	**6,428**	**(18,346)**
Segment results – adjusted EBITDA				
Bronchitol & Aridol	(1,198)	(2,421)	(3,786)	(7,100)
New drug development	(3,683)	199	28,771	(4,114)
Corporate	(947)	(1,005)	(13,466)	(4,017)
Total	**(5,827)**	**(3,227)**	**11,519**	**(15,231)**
Statement of cash flows				
Cash inflow/ (outflow) from:				
Operations	(2,712)	(4,228)	12,206	(15,262)
Investing activities	(280)	(328)	(884)	(723)
Financing activities	(443)	(434)	(1,753)	(1,721)
Total cash generated/(used)	**(3,435)**	**(4,990)**	**9,569**	**(17,706)**
Cash at bank	**31,073**	**21,504**	**31,073**	**21,504**

Highlights for the quarter

- In comparing the revenue for the quarter with the prior comparable period please note:
  - The growth in sales was mainly attributable to increased sales to Chiesi for the UK and German markets.
  - Total revenue in the comparable 2017 quarter included recognition of an R&D tax credit of $3.2 million and clinical trial cost reimbursements of $2.6 million. The Company does not qualify for an R&D tax credit in 2018 due to total revenue for the year exceeding the $20 million cap. The clinical trial to which the reimbursement related was substantially completed in 2017.
- Expenses
  - Total expenses decreased for the quarter, driven mainly by two line items
    - Clinical trial expenses in relation to clinical trial CF303 changed from $3.2 million in the June 2017 quarter to a credit of $30,000 in the June 2018 quarter. Clinical trial expenses in the current quarter relate to the two phase 1 trials being run as part of the LOXL2 program.
    - New drug development expenses decreased from $2.2 million in the June 2017 quarter to $1.6 million in the current quarter. In 2017 these costs related mainly to the Company's LOXL2 program, while in the current quarter the costs related mainly to the

other pipeline projects, specifically the oral LOX program and the SSAO/MPO program which are progressing towards human clinical trials.
    - Note that year to date expenses include $9.6 million of costs incurred in the December quarter associated with changes to the collaboration agreement with Synairgen.
  - Foreign exchange gains and losses include an unrealised gain in the comparative quarter of $0.1 million and an unrealised loss in the current quarter of $817,000.
  - Other expenses for the quarter include $235,000 (a credit of $90,000 in the comparative quarter) representing 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, as well as other inventory adjustments.
- The closing cash position at 30 June 2018 was $31 million.

Segment information

A$'000	Segment information - three months ended							
(unaudited)	30-June-18				30-June-17			
Income statements	Bronchitol & Aridol	New drug developm't	Corporate	Total	Bronchitol & Aridol	New drug developm't	Corporate	Total
Revenue								
Sale of Bronchitol	1,291	-	-	1,291	277	-	-	277
Sale of Aridol	609	-	-	609	589	-	-	589
	1,900	**-**	**-**	**1,900**	**866**	**-**	**-**	**866**
Milestones from sale of drug	-	-	-	-	-	-	-	-
Clinical reimbursement	-	-	-	-	2,592	-	-	2,592
Tax credit	-	-	-	0	54	3,045	-	3,099
Other revenue	5	-	122	128	168	-	86	254
	1,905	**-**	**122**	**2,028**	**3,680**	**3,045**	**86**	**6,811**
Expenses								
Employee costs	(1,490)	(722)	(442)	(2,654)	(1,581)	(512)	(530)	(2,623)
Clinical trials	30	(1,175)	-	(1,145)	(3,210)	-	-	(3,210)
Drug discovery	-	(1,589)	-	(1,589)	-	(2,165)	-	(2,165)
Other expenses	(1,643)	(197)	(627)	(2,467)	(1,310)	(169)	(561)	(2,040)
Total expenses	(3,103)	(3,683)	(1,069)	(7,855)	(6,101)	(2,846)	(1,091)	(10,038)
Adjusted EBITDA	**($1,198)**	**($3,683)**	**($947)**	**($5,827)**	**($2,421)**	**$199**	**($1,005)**	**($3,227)**

Commentary for the quarter

- Bronchitol & Aridol:
  - Sales of Bronchitol and Aridol increased as detailed in commentary above.
  - Clinical trial reimbursements and clinical trial costs ceased following completion of study CF303 in 2017.
  - The reduction in employee costs relates to clinical employees now predominantly focused on the New Drug Development programs.
  - Other expense increases include higher manufacturing costs associated with higher sales production volumes and also inventory adjustments.
- New drug development:
  - As noted above the Company does not qualify for an R&D tax credit in 2018 due to total revenue for the year exceeding the $20 million cap. In 2017 the Company booked an R&D tax credit of $3 million in relation to New Drug Development.
  - Clinical trial expenses relate to the 2 phase 1 trials being conducted in the LOXL2 program.

- Drug discovery expenses include work on the LOXL2 program ($227,000 for the quarter; $1.4 million in 2017), and the two programs currently in preclinical studies - the LOX program ($398,000 for the quarter; $222,000 in 2017) and the SSAO/MPO program ($787,000 for the quarter; $382,000 in 2017).

A$'000	Segment information - twelve months ended							
(unaudited)	30-June-18				30-June-17			
Income statements	Bronchitol & Aridol	New drug developm't	Corporate	Total	Bronchitol & Aridol	New drug developm't	Corporate	Total
Revenue								
Sale of Bronchitol	4,084	-	-	4,084	2,785			2,785
Sale of Aridol	2,010	-	-	2,010	2,038			2,038
	6,094	**-**	**-**	**6,094**	**4,823**	**-**	**-**	**4,823**
Milestones from sale of drug	-	42,130	-	42,130	-	-	-	-
Clinical reimbursement	1,188	-	-	1,188	8,463	-	-	8,463
Tax credit	-	161	-	161	70	3,089	-	3,159
Other revenue	186	5	471	662	188	328	336	853
	7,468	**42,296**	**471**	**50,235**	**13,544**	**3,417**	**336**	**17,298**
Expenses								
Employee costs	(5,695)	(2,753)	(1,883)	(10,331)	(6,037)	(2,026)	(2,058)	(10,121)
Clinical trials	(160)	(3,465)	-	(3,625)	(10,017)	-	-	(10,017)
Drug discovery	-	(6,816)	-	(6,816)		(5,068)	-	(5,068)
Other expenses	(5,399)	(491)	(2,474)	(8,364)	(4,590)	(437)	(2,295)	(7,322)
Change in collaboration	-	-	(9,580)	(9,580)	-	-	-	-
Total expenses	(11,254)	(13,525)	(13,937)	(38,716)	(20,644)	(7,531)	(4,353)	(32,528)
Adjusted EBITDA	**($3,786)**	**$28,771**	**($13,466)**	**$11,519**	**($7,100)**	**($4,114)**	**($4,017)**	**($15,230)**

Commentary for the twelve months

- Bronchitol & Aridol:
  - Sales of Bronchitol increased for Australia (27% to $949,000); Western Europe (157% to $2.9 million) including sales to Chiesi for Germany, Italy and the UK; while the comparative period includes a sale of $643,000 for Russia where the next sale is expected later in the 2018 calendar year. Bronchitol sales to Eastern Europe (predominantly Turkey and the Czech Republic) for the year were $260,000 compared to $278,000 in 2017, and the current year includes the first sale to the Company's distributor in the Czech Republic ($129,000)
  - Clinical trial reimbursements and clinical trial costs reduced following reporting of study CF303 in June 2017.
  - The reduction in employee costs relates to clinical employees now predominantly focused on the New Drug Development programs.
  - The increase in Other expenses is primarily due to higher manufacturing costs associated with higher sales production volumes, the net transfer of labour and overhead into and inventory adjustments as discussed above.
- New drug development:
  - During 2018 the Company received two milestone payments from Boehringer Ingelheim as the drug candidate they acquired in 2015 commenced phase 2a clinical trials in two separate disease indications.

- As noted above the Company does not qualify for an R&D tax credit in 2018 due to total revenue for the year exceeding the $20 million cap. In 2017 the Company booked an R&D tax credit of $3 million in relation to New Drug Development.
- Increased new drug development expenses for the three quarters reflects
  - Staff increases earlier in the financial year
  - The commencement of 2 phase 1 clinical trials in relation to the LOXL2 program
  - Drug discovery expenditure for the year includes the LOXL2 program ($2.3 compared to $3.0 million in 2017); the SSAO/MPO program ($2.0 million compared to $1.0 million in 2017) and the LOX program ($1.9 million compared to $0.6 million in 2017).

- Corporate:
  - Note the $9.6 million of costs incurred in the December quarter associated with changes to the collaboration agreement with Synairgen.

Income statements

A$'000	Three months ended		Twelve months ended	
(unaudited)	**30-June-18**	**30-June-17**	**30-June-18**	**30-June-17**
Revenue				
Revenue from sale of goods	1,900	866	6,094	4,823
Milestones from sale of drug	-	-	42,130	-
Clinical trial cost reimbursements	1	2,592	1,188	8,463
Interest	185	135	598	703
Drug discovery service fee	-	(2)	-	328
R&D tax incentive	161	3,160	161	3,160
Other	(34)	194	662	524
Total revenue	**$2,213**	**$6,945**	**$ 50,833**	**$ 18,001**
Expenses				
Employee costs	(2,949)	(2,860)	(11,531)	(11,063)
Administration & corporate	(488)	(549)	(2,310)	(1,947)
Rent, occupancy & utilities	(349)	(303)	(1,279)	(1,148)
Clinical trials	(1,145)	(3,210)	(3,625)	(10,017)
Drug development	(1,589)	(2,165)	(6,816)	(5,068)
Sales, marketing & distribution	(266)	(346)	(1,163)	(1,061)
Safety, medical and regulatory affairs	(312)	(430)	(885)	(1,379)
Manufacturing purchases	(533)	(311)	(1,774)	(1,326)
Other	(519)	(201)	(1,134)	(437)
Depreciation & amortisation	(760)	(768)	(3,112)	(3,059)
Foreign currency exchange gains & losses	(817)	232	(641)	781
Finance costs	(132)	(146)	(563)	(623)
Costs in relation to change in collaboration agreement	-	-	(9,580)	-
Total expenses	**(9,859)**	**(11,057)**	**(44,413)**	**(36,347)**
Net profit (loss) before tax	**(7,646)**	**(4,112)**	**6,420**	**(18,346)**
Income tax credit/(expense)	-	-	8	-
Net profit (loss) after tax	**($7,646)**	**($4,112)**	**$6,428**	**($18,346)**

Summary balance sheets

A$'000		
(unaudited)	**30-June-18**	**30-June-17**
Assets		
Cash	31,073	21,504
R&D tax credit receivable	-	3,100
Accounts receivable	1,787	1,262
PP&E	12,451	14,860
Other	4,786	4,708
	$50,097	**$45,434**
Liabilities		
Accounts payable and accrued expenses	4,926	6,134
Lease liability (Frenchs Forest facility)	8,268	9,251
Financing agreement (not repayable other than as a % of US & EU Bronchitol revenue)	22,754	22,141
Other liabilities	3,031	4,387
	$38,979	**$41,913**
Net Assets	**$11,118**	**$3,521**