

Quarterly Shareholder Update – June 2016

Pharmaxis completes first year of new business plan



Dear Shareholder,

June 30, 2016 marks the completion of Pharmaxis' first year of operation following a business restructure which concluded with the sale of our drug candidate PXS-4728A to Boehringer Ingelheim (Boehringer) in May of 2015. Progress under the new business model over the 2016 financial year has been substantial.

Importantly, Boehringer's most recent status report on the development program for PXS-4728A has again confirmed its expectation that a phase 2 trial of PXS-4728A will commence in the first quarter of 2017, at which time a milestone payment of approximately A\$25 million will be payable to Pharmaxis. As I have made clear over the last year, I believe Boehringer to be a perfect partner for this drug. It is a company with an extensive metabolic franchise and a clear belief in SSAO as a well validated target in NASH – an increasingly common disease with a high unmet need and a huge potential market. Our drug is Boehringer's lead clinical candidate for this new and emerging market so there is a great strategic fit that I expect will benefit Pharmaxis and its shareholders for many years.

Our drug discovery program to develop new drugs for fibrotic diseases such as NASH and pulmonary fibrosis (IPF) has delivered several very suitable drug candidates that we will now assess in conjunction with our UK collaborator Synairgen to determine which should be progressed into human clinical trials in 2017. The search for good anti fibrotic medications remains highly competitive and the once-a-day oral small molecule inhibitors of the enzyme LOXL2 produced by our in house program offer great hope for patients. Interest in this program from large pharmaceutical companies with whom we have an ongoing dialogue remains strong and is actively shaping our pre-clinical development program.

Our other drug discovery programs have also made substantial progress. In particular the neuro inflammation program where we have identified a lead compound and have started assessing the appropriate disease indication for further development based on feedback from potential partners.

Finally, two weeks ago we announced the completion of recruitment for our phase 3 Bronchitol clinical trial in cystic fibrosis. That trial is designed to meet the remaining clinical requirements of the complete response letter received from the US Food and Drug Administration (FDA) in 2013. We expect to receive the results of the trial in the second quarter of 2017 and assuming a positive outcome, the submission of the trial results to the FDA by Chiesi later that year.

The Company finished the financial year with a cash balance of \$39 million and net cash usage over the year of \$15 million. We are well positioned as we move into a particularly exciting time.

This report outlines our recent progress.

Sincerely,

A handwritten signature in black ink that reads "Gary Phillips". The signature is written in a cursive, flowing style with a long horizontal stroke extending to the right.

Chief Executive Officer

Drug discovery

Boehringer Ingelheim advancing PXS-4728A towards a phase 2 clinical trial

Boehringer acquired PXS-4728A in May 2015 to develop initially as a treatment for non-alcoholic steatohepatitis (NASH). Under the terms of our agreement, Boehringer has total responsibility for the development program and is required to make milestone payments to Pharmaxis as PXS-4728A progresses towards approval as well as other sales related payments post approval.

Boehringer is currently completing the design of the phase 2 clinical program and the prerequisite toxicology studies, manufacturing scale up and drug formulation. Pharmaxis received a periodic progress report from Boehringer in late June in which it again confirmed that PXS-4728A will commence a phase 2 NASH trial in the first quarter of 2017, triggering a milestone payment to Pharmaxis of approximately A\$25 million.

Pharmaxis is also entitled to a milestone payment should Boehringer commence a phase 2 trial in a second indication.

Boehringer is a global leader in pharmaceutical products for cardiometabolic diseases and the NASH indication is a very large unmet need in this area being pursued by many pharmaceutical companies. Boehringer is therefore a strong and strategically aligned partner to develop PXS-4728A and we continue to be very pleased with their commitment to the development program.

Chemistry for LOXL2 inhibitor program now complete

Pharmaxis is developing selective inhibitors to the lysyl oxidase type 2 enzyme (LOXL2) utilising the amine oxidase platform that delivered PXS-4728A. The Company is focusing its efforts on NASH and kidney fibrosis and is collaborating with UK biotechnology company Synairgen plc (LSE: SNG) to develop a LOXL2 inhibitor to treat the fatal lung disease idiopathic pulmonary fibrosis (IPF). The LOXL2 enzyme also plays a role in some solid cancers.

The Pharmaxis drug discovery group has developed a small number of LOXL2 inhibitors that are suitable for further development. The lead optimisation work of the chemistry team is complete and together with Synairgen we will now focus on selecting which drug candidates to take forward into the clinic. We are on track to select a LOXL2 drug candidate for IPF and/or NASH and proceed into full preclinical evaluation in the second half of 2016, and are targeting commencement of a phase 1 clinical trial in 2017.

Pharmaceutical company interest continues in the LOXL2 inhibitor program

There is a continued very high interest among pharmaceutical companies in new drugs that inhibit the LOXL2 enzyme due to the role it is believed to play in a number of fibrotic diseases including NASH, the fatal lung disease idiopathic pulmonary fibrosis (IPF), kidney fibrosis and cardiac fibrosis. The deal values for phase 1 and phase 2 transactions in the fibrosis space generally are very significant.

Pharmaxis CEO Gary Phillips met with over a dozen companies at the recent international BIO conference in San Francisco to update them on our LOXL2 research program and better understand the objectives and data requirements of these potential partners. A special Shareholder Update on the BIO conference was released on 21 June and is available on the [Pharmaxis website](#).

We will continue to interact with these larger companies as our development program progresses and expect that our LOXL2 small molecule inhibitor program will generate a competitive partnering process after completing phase 1 clinical trials, currently scheduled to commence in the second half of 2017.

Drug development pipeline – other programs

The recent international BIO conference was also an opportunity to discuss our neuro inflammation SSAO/MAOB program with pharmaceutical companies. We identified a number of companies that see a strong need for a centrally acting non-steroidal anti-inflammatory for neuro inflammation and other indications. We have a

lead candidate for this program and will now focus on selecting the appropriate indication before proceeding to formal preclinical development and phase 1 studies.

As reported last quarter we have research collaborations with a number of leading universities and academics assessing the utility of our LOXL2 inhibitors in oral cancer, bone marrow myelofibrosis, NASH and wound scarring.

Pharmaxis establishes Scientific Advisory Board

Pharmaxis has established a scientific advisory board to overview the Company's drug discovery and development programs. The Company will expand from this initial core group as required with other invited members with knowledge and expertise in developing assets in fibrosis and inflammation. The scientific advisory board augments the key opinion leaders, academics and industry participants with whom Pharmaxis engages in all of its individual programs.

The initial two members of the scientific advisory board are Professor Jacob George and Dr Alan Robertson.

Professor Jacob George is the Robert W Storr Professor of Hepatic Medicine at the Storr Liver Unit, Westmead Millennium Institute, University of Sydney and is Head of the Department of Gastroenterology and Hepatology at Westmead Hospital and Director of Gastroenterology and Hepatology Services for the Western Sydney Local Health Network. He undertakes basic and clinical research on hepatitis C, liver cancer, NASH and hepatic fibrosis.

Dr. Alan Duncan Robertson, B.Sc., Ph.D. served at Wellcome plc in London, Faulding Ltd, Amrad Ltd and was chief executive officer of Pharmaxis Ltd from December 1999 to March 2013. He is the co-inventor of 18 patents and author of more than 35 scientific papers. Dr Robertson is also the inventor of the migraine therapeutic Zomig, which is marketed worldwide by AstraZeneca.

Further details are available on the [Pharmaxis website](#).

Bronchitol for cystic fibrosis

Bronchitol[®] is an inhaled dry powder for the treatment of cystic fibrosis and has been the subject of two large scale global clinical trials conducted by Pharmaxis. The product is approved and marketed in Europe and Australia and a third large multicentre clinical trial is currently underway aiming to secure approval in the United States.

United States

In the US Pharmaxis has partnered with Chiesi Farmaceutici SpA (Chiesi) to conduct the international phase 3 clinical trial (CF303) designed to meet the remaining clinical requirements of the US Food and Drug Administration (FDA). Under the terms of the agreement and following a positive outcome of the trial, Chiesi will have responsibility for completing the New Drug Application with the FDA and the commercialisation of Bronchitol in the United States. We continue to work closely with Chiesi on all aspects of securing US marketing approval for Bronchitol.

On 15 July we were pleased to announce the completion of recruitment for the clinical trial which commenced recruitment in October 2014. It is being conducted in 126 sites across 21 countries and, subject to final randomisation of patients screened by the trial sites, the final enrollment is expected to reach 420 adult CF patients. The results of the trial are expected to be reported in the second quarter of 2017. Subject to a positive trial outcome, Chiesi will submit a response to the FDA and a decision on approval can be expected in 2018.

Chiesi is responsible for funding up to US\$22 million of the cost of the trial, the total cost of which is expected to be approximately US\$26 million. Milestones totaling up to US\$25 million are payable to Pharmaxis including US\$10 million on the launch of Bronchitol.

Europe

In the EU, Pharmaxis appointed Chiesi as its exclusive distributor for the currently launched markets of the UK and Germany from 1 June 2015. Chiesi is an experienced and respected partner in key global markets and sells Bronchitol as part of its cystic fibrosis portfolio.

Chiesi did not purchase additional Bronchitol during the June quarter having built inventory levels over prior periods in its central European warehouse and at its wholesalers. Available in-market sales data to 31 March indicates total sales slightly below the comparative quarter which we consider a satisfactory outcome given the change in distribution to Chiesi in 2015 and significant Chiesi marketing initiatives only commencing in 2016. Ongoing Bronchitol sales by Pharmaxis will continue to reflect the timing of Chiesi orders to replenish inventory rather than in market use of the product.

CF204 presented at European Cystic Fibrosis Conference

The results of Pharmaxis phase 2 trial of Bronchitol in children and adolescents with cystic fibrosis (CF204) were presented at the annual European Cystic Fibrosis Conference held in Basel, Switzerland from the 8th to 11th June 2016. Prof Dr Kris De Boeck, President of the European Cystic Fibrosis Society Board made an oral presentation and concluded:

“In this randomised, placebo-controlled crossover trial in subjects with CF aged 6-17 years, inhaled mannitol was associated with significant improvements in both primary and secondary end points. The study confirms that inhaled mannitol as add-on therapy to optimal care provides immediate and significant benefits in lung function and sputum weight in children and adolescents. These improvements were seen on top of usual treatments and irrespective of rhDNase use, age or disease severity. Inhaled mannitol was not associated with a greater safety risk than that observed with placebo and serious adverse events were less common in the mannitol group. Inhaled mannitol was associated with a reduced incidence of pulmonary exacerbations and the incidence of haemoptysis was low and

similar between groups. (Clinical Trials.Gov: NCT 01883531)”

Given these results are on top of the current standard of care and are seen to be clinically significant, the Company has determined to seek an extension of the EU marketing authorisation to include children and adolescents and in accordance with mandated regulatory timeframes will submit an application in early 2017. It is not yet known if the trial results alone will be sufficient to gain approval of an extended label.

Other territories

Approval and reimbursement applications continue to progress in various countries including Russia, Eastern Europe, the Middle East and Brazil. Russian approval is expected shortly.

Corporate

Pharmaxis in the media

Australia’s independent research publication Bioshares interviewed Gary Phillips both before and after his attendance at the 2016 international BIO conference in San Francisco in June 2016. The Bioshares articles are available on the [Pharmaxis website](#).

Pharmaxis corporate profile

A two page overview of the Pharmaxis business, catalysts and risks was updated during the quarter and is available on the [Company website](#).

Updated investor presentation

In conjunction with international and Australian investor meetings an updated company presentation in early June and is also available on the [Pharmaxis website](#).

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Financials

Highlights:

- Sales revenue for the quarter was lower than the comparable period as discussed above. However, sales revenue for the full year was maintained despite the appointment of Chiesi as a distributor in two major markets at the end of the 2015 financial year resulting in a share of ongoing sales revenue being retained by the distributor.
- Other revenue in 2015 included the sale of drug candidate PXS-4728A to Boehringer for \$41 million in upfront payments. The next milestone is expected in the first quarter of calendar 2017.
- Other revenue in the 2016 year included an R&D tax incentive of \$2.1 million which was recorded in the June quarter.
- With the exception of clinical trial costs which predominantly related to CF303 (which is reimbursed by Chiesi up to US\$22 million and included in other revenue) and drug discovery costs where the Company is investing, core operating expenses have reduced significantly.
- Segment information on page 9 provides a useful overview of the business.
- The closing cash of \$39 million and the reduced net annual cash usage places the Company in a strong position.

Key financial metrics for the period are as follows:

	A\$'000	Three months ended		Twelve months ended	
		30-June-16	30-June-15	30-June-16	30-June-15
(unaudited)					
Income statements					
Sales		703	1,645	6,135	5,999
Total revenue		4,794	42,712	19,020	59,247
Total expenses		(6,919)	(12,126)	(35,476)	(40,739)
Net profit (loss) after tax		(2,125)	30,639	(16,463)	18,466
Segment results – adjusted EBITDA					
Bronchitol & Aridol		(2,140)	(4,187)	(8,228)	(10,045)
New drug development		92	36,159	(2,625)	35,068
Corporate		(1,133)	(703)	(3,988)	(3,532)
Total		(3,181)	31,269	(14,841)	21,491
Statement of cash flows					
Cash inflow/ (outflow) from:					
Operations		(1,725)	31,428	(12,155)	21,780
Investing activities		(147)	(114)	(1,382)	(264)
Financing activities		(425)	(300)	(1,714)	(1,791)
Total cash used		(2,297)	31,014	(15,251)	19,725
Foreign currency exchange rate changes impact on cash		(2)	118	322	231
Cash at bank		39,209	54,138	39,209	54,138

Income statements for the quarter and year and supplementary commentary are below. Additional financial information will be available in the Preliminary Final Statement scheduled to be released on 19 August 2016.

Income statements:

1. Sales revenue is summarised in the Segment Information attached.
 - Bronchitol sales in our largest launched markets, Germany and the United Kingdom, represent approximately 80% of total Bronchitol sales. Bronchitol was sold by Pharmaxis directly to pharmacies in these markets until 31 May 2015 after which Bronchitol was sold via our exclusive distributor Chiesi. Sales by Pharmaxis to Chiesi are at a lower unit price to allow for distributor margins. As noted above, Chiesi did not purchase additional Bronchitol during the June 2016 quarter having built inventory levels over prior periods in its central European warehouse and regional wholesalers. Ongoing quarterly Bronchitol sales by Pharmaxis will continue to reflect the timing of distributor orders to replenish inventory rather than in market use of the product.
 - Aridol sales increased in both the quarter and the year, with growth seen in all markets from increased unit sales and/or pricing.
2. Other revenue and income as detailed on the attached Income Statements discloses:
 - Clinical trial cost reimbursements by our US partner Chiesi in relation to the ongoing phase 3 clinical trial of Bronchitol. Under our agreement, Chiesi is responsible for the first US\$22 million of costs. The revenue recognised each period represents clinical trial costs invoiced to Chiesi reduced by a revenue deferral designed to recognise Pharmaxis' expected funding requirement at the end of the trial (currently estimated at up to US\$ 4 million) over the term of the trial. The total deferred revenue at 30 June 2016 is A\$3.7 million, A\$ 1.1 million of which was deferred in the June 2016 quarter and A\$2.7 million in total for the 2016 financial year. With the closing of recruitment in July 2016 the total cost of the trial is not expected to increase substantially from current estimates. Note that approximately \$3.2 million of the \$11.1 million recorded as income in the 2015 financial year was reimbursement for costs incurred and expensed in the 2014 financial year, reimbursed by Chiesi after the agreement was signed in December 2014.
 - The sale of drug candidate PXS-4728A in May 2015 to Boehringer Ingelheim for A\$41 million.
 - An R&D tax credit in relation to the 2016 financial year of \$2.1 million. The Company was not eligible for the R&D tax credit in the 2015 financial year.
 - The drug discovery service fee is amounts charged to Synairgen under our research collaboration agreement.
3. Employee costs decreased in line with the reduced number of employees when compared with the prior periods.
4. Clinical trial costs for the quarter relate to the phase 3 clinical trial in cystic fibrosis and are reimbursed by Chiesi up to US\$22 million, as described above. Costs in relation to the phase 2 paediatric trial conducted in Europe that completed and reported during the December were \$Nil for the June 2016 quarter and \$645,000 for the 2016 financial year. Clinical trial costs also include the phase 1 trial for drug candidate PXS-4728A which was substantially completed in the 2015 financial year – as shown in the attached Segment Information.
5. Drug discovery costs have increased in line with the Company's increased focus on developing new drugs from its amine oxidase chemistry platform.
6. Other expenses include the net transfer of manufacturing labour and overhead into inventory which in the current quarter was greater than other expenses incurred.

7. Foreign currency exchange gains and losses includes an unrealised loss for the June quarter of \$885,000 (2015: \$149,000 gain) and for the 2016 financial year an unrealised loss of \$911,000 (2015: \$1,495,000 loss) in relation to the financing agreement with NovaQuest.
8. Finance expenses relates to the financing agreement with NovaQuest and the Company's finance lease for its Frenchs Forest facility. In the current quarter the Company revised the assumptions on which the financing liability is calculated including the quantum and timing of forecast sales on which future payments are expected to be made and expected foreign currency rates used to forecast sales. As a result the liability was reduced by A\$3.1 million with a corresponding reduction in finance expense. In the 2015 financial year the Company completed an amended financing agreement which resulted in a negative finance expense for the year of \$2.7 million.

Statement of cash flows:

9. Cash flow used in operations in the prior period included an R&D tax incentive of \$3.4 million relating to the 2014 financial year. The Company was not eligible for the incentive in the 2015 financial year. The 2016 R&D tax incentive of \$2.1 million is expected to be received in the second half of calendar 2016 after filing of the Company's 2016 income tax return.
10. Investing activities for the year predominantly relate to manufacturing cost reduction initiatives, the replacement of IT infrastructure and new analytical equipment for drug discovery.

Income statements

	A\$'000	Three months ended		Twelve months ended	
		30-June-16	30-June-15	30-June-16	30-June-15
(unaudited)					
Income statements					
Revenue					
Revenue from sale of goods		703	1,645	6,135	5,999
Sale of drug candidate		-	38,814		40,603
Clinical trial cost reimbursements		1,249	1,966	8,200	11,139
Interest		298	203	1,213	721
R&D tax incentive		2,100	-	2,100	164
Drug discovery service fee		257		925	-
Other		187	84	447	621
Total revenue		4,794	42,712	19,020	59,247
Expenses					
Employee costs		(2,862)	(3,099)	(10,529)	(14,111)
Administration & corporate		(531)	(799)	(2,082)	(3,316)
Rent, occupancy & utilities		(321)	(402)	(1,296)	(1,593)
Clinical trials		(2,453)	(4,418)	(11,955)	(11,315)
Drug development		(1,138)	(878)	(2,910)	(1,695)
Sales, marketing & distribution		(246)	(349)	(1,101)	(1,962)
Safety, medical and regulatory affairs		(344)	(631)	(1,707)	(1,723)
Manufacturing purchases		(785)	(395)	(1,928)	(1,737)
Other		553	(1,154)	(381)	(1,905)
Foreign currency exchange gains & losses		(850)	(845)	(844)	(395)
Depreciation & amortisation		(755)	(738)	(3,028)	(3,406)
Finance expenses		2,975	108	2,459	2,696
Impairment expense		(174)	(277)	(174)	(277)
Total expenses		(6,919)	(12,126)	(35,476)	(40,739)
Net profit (loss) before tax		(2,125)	30,586	(16,456)	18,508
Income tax expense		-	53	(7)	(42)
Net profit (loss) after tax		(2,125)	30,639	(16,463)	18,466

Segment information

A\$'000	Segment information - three months ended							
(unaudited)	30-June-16				30-June-15			
Income statements	Bronchitol & Aridol	New drug developmt	Corporate	Total	Bronchitol & Aridol	New drug developmt	Corporate	Total
Revenue								
Sale of Bronchitol	187	-	-	187	1,241	-	-	1,241
Sale of Aridol & other	516	-	-	516	405	-	-	405
	703	-	-	703	1,646	-	-	1,646
Sale of drug candidate	-	-	-	-	-	38,814	-	38,814
Clinical reimbursement	1,249	-	-	1,249	1,966	-	-	1,966
Tax credit	544	1,556	-	2,100	-	-	-	-
Other revenue	30	356	58	444	-	-	84	84
	2,526	1,912	58	4,496	3,612	38,814	84	42,510
Expenses								
Employee costs	(1,348)	(530)	(579)	(2,457)	(1,698)	(651)	(976)	(3,325)
Clinical trials	(2,453)	-	-	(2,453)	(3,410)	(1,007)	-	(4,417)
Drug development	-	(1,138)	-	(1,138)	-	(878)	-	(878)
Other expenses	(865)	(152)	(612)	(1,629)	(2,691)	(119)	189	(2,621)
Total expenses	(4,666)	(1,820)	(1,191)	(7,677)	(7,799)	(2,655)	(787)	(11,241)
Adjusted EBITDA	(2,140)	92	(1,133)	(3,181)	(4,187)	36,159	(703)	31,269
A\$'000	Segment information - twelve months ended							
(unaudited)	30-June-16				30-June-15			
Income statements	Bronchitol & Aridol	New drug developmt	Corporate	Total	Bronchitol & Aridol	New drug developmt	Corporate	Total
Revenue								
Sale of Bronchitol	4,302	-	-	4,302	4,243	-	-	4,243
Sale of Aridol & other	1,833	-	-	1,833	1,756	-	-	1,756
	6,135	-	-	6,135	5,999	-	-	5,999
Sale of drug candidate	-	-	-	-	-	40,603	-	40,603
Clinical reimbursement	8,200	-	-	8,200	11,139	-	-	11,139
Tax credit	544	1,556	-	2,100	164	-	-	164
Other revenue	31	1,024	317	1,372	239	-	382	621
	14,910	2,580	317	17,807	17,541	40,603	382	58,526
Expenses								
Employee costs	(5,560)	(1,782)	(2,116)	(9,458)	(9,615)	(1,692)	(2,613)	(13,920)
Clinical trials	(11,846)	(109)	-	(11,955)	(9,469)	(1,846)	-	(11,315)
Drug development	-	(2,910)	-	(2,910)	-	(1,695)	-	(1,695)
Other expenses	(5,732)	(404)	(2,189)	(8,325)	(8,502)	(302)	(1,301)	(10,105)
Total expenses	(23,138)	(5,205)	(4,305)	(32,648)	(27,586)	(5,535)	(3,914)	(37,035)
Adjusted EBITDA	(8,228)	(2,625)	(3,988)	(14,841)	(10,045)	35,068	(3,532)	(21,491)