

THE ACHIEVER

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AWESOME AUTUMN NEWS

- Update on Australian Inherited Retinal Disease Register and DNA Bank, and how to participate
- Update on 2009 Retina Australia Congress in Brisbane
- Inspirational Human Interest Story
- Research Developments



PP: 33 1088/00015

From the President

Since the previous edition of the Achiever, we have had a very busy and productive time in and around the office. Our administrative officer, Mary Maga, has left us after fourteen months to pursue other interests. Mary's contribution was greatly appreciated and we wish her all the best with her future endeavours. The Board is currently looking for an administration or finance manager and are confident that a suitable replacement will be found in the very near future.

The Telelink, which was mentioned in the last newsletter, has commenced and already has proven to be a great success. There are eleven participants who come together by phone each Tuesday from Warrnambool, Inverloch, Heyfield, Shepparton, Traralgon, Albury, Eden Park, Werribee, Patterson Lakes, Melbourne and Launceston. I am extremely grateful to Rose Petrotta who has willingly taken on the role as host of this project. As I mentioned last time, if this trial telelink is successful, we may be able to expand the project to include other interested members.

As you are all probably aware, the 2009 Easter Raffle has commenced. I would like to encourage you all to purchase, or sell, tickets returning them to the office in time for the draw on Tuesday 7 April. We have had some wonderful prizes donated, hence all of the monies raised will be able to go directly to research. At the time of writing this article, we have raised in excess of \$1000 from ticket sales. Please keep the money coming in, every dollar helps. If you need additional books, please phone the office and they will be sent to you as quickly as possible.

I encourage you to attend our Easter Luncheon which will be held after the raffle draw, at 12.30pm on Tuesday 7 April. We are really looking forward to making this a wonderful celebration and time to catch up with members. If you are interested in attending, it is important that you RSVP as soon as practicable, by phoning the office on 9650 5088, so that we can make an appropriate booking. We are planning to book "The Spaghetti Tree" at 59 Bourke Street, Melbourne, so please confirm your numbers soon.

On Sunday 1 March, a number of volunteers spent the day representing Retina Australia Vic at the Guide Dogs of Victoria's Open Day. We erected our marquee and spent the day handing out brochures, tunnel vision specs and other leaflets of information about retinal diseases. This day was most successful in raising public awareness of our organisation as well as reaching out to potential members.

Finally, I would like to draw your attention to the Congress information which is included in this edition. I encourage you to think about attending this event in October, as the program is coming together very well and includes some excellent speakers who will tell us about the latest research across the globe.

Leighton Boyd

FEATURES

In the President's Report of the last edition of The Achiever, Leighton briefly mentioned an exciting new Australian initiative supported by Retina Australia to assist researchers in finding a cure for inherited retinal diseases through the development of a national inherited retinal disease register including the collection of DNA samples for genetic analysis. In this edition, Leighton provides further details below. This is followed by a WA government media release about this project.

BREAKING NEWS

YOUR CHANCE TO BE INVOLVED

AUSTRALIAN INHERITED RETINAL DISEASE REGISTER AND DNA BANK PROGRAM

Throughout recent years members will have read of research work being carried out with retinal gene therapies, firstly with animal models, both overseas and here in Australia, and during the past couple of years, clinical trials have commenced involving persons affected by inherited retinal diseases (IRDs). The very significant success with the animal trials gave the impetus for the human trials to be approved and to commence.

In many cases, the development and application of a gene therapy for an individual requires knowledge of the nature of that individual's genetic defect. In preparation for potential gene therapies and possible treatment, a person's errant gene type needs to be known – this is done by provision of a blood sample (or in some instances, particularly for young and elderly people a saliva sample) from which the DNA is extracted and then 'sequenced' to determine the specific gene responsible for the IRD.

For some time Retina Australia has recognised the need for an Australian IRD Register and DNA Bank aimed at determining the gene status of individuals. Ideally, in due course, all persons with an IRD and nominated blood relatives, if they so wish, would have their retinal gene status determined. It has been accepted that there is a need for early actions in developing the Program given that Prof Robin Ali, the leader of the human retinal disease gene therapy trials at Moorfields Hospital in London, indicated at the recent Retina International Congress in Helsinki, that gene therapies are expected to become available in the UK within the next 3 years to persons diagnosed with the specific IRD gene, Leber's Congenital Amaurosis.

An IRD Register and DNA Bank, funded by the Western Australian Retinitis Pigmentosa Foundation (WARPF), has been operating in that state for the past

several years utilising facilities at the Sir Charles Gairdner Hospital in Perth. The other State members of Retina Australia have agreed to fund an extension of the program for the rest of Australia. We are proud to say that this program will commence on 1 April, 2009.

As alluded to above, involvement in the program is voluntary however all members are strongly urged to participate. An Expression of Interest form has been mailed to all members, including those who normally receive email or audio newsletters, in a special mail out. If you are interested in personally being involved in this Australian research, you will need to complete this form, sign it and then send the form to the nominated Department at the Perth Hospital. In due course the a member of the project team will then contact the member seeking family history information, including a family tree covering blood relatives.

The Project team will send all of the necessary documents to every individual who has expressed interest in having their gene type identified from a blood or saliva sample. This will include information, forms, return envelopes, a letter addressed to a pathologist local to the person and the required materials for use by the pathologist in collecting the sample. Other than the cost of getting to and from the pathologist there will be no cost to the program participants – such costs being covered by the Retina Australia funding.

For privacy, and possible family sensitivity reasons, the results of the DNA extraction, and subsequent analysis, will not be made known to the individual participants. However, if specific results show that the participant could possibly be eligible for gene therapies, as they become available, the participant will be advised to make contact with a genetic counsellor to whom the DNA information will be made available in order to discuss their personal involvement and possible treatment. Some participants may be invited to become directly involved with the gene therapy trials.

At the Retina International Congress mentioned previously, it was indicated that gene therapy trials are in the pipeline for specific IRD genes or genetic groups including Leber's Congenital Amaurosis, Stargardt Disease, Ushers (particularly Type 3) and Achromatopsia. It is generally accepted that gene therapy activities are no longer regarded as solely research related activities, they are now close to being part of the clinical diagnosis and treatment process.

This further emphasises the vital need for the Australian IRD Register and DNA Bank program to be underway on 1 April, 2009. If you wish to become involved, so that your blood or saliva can be tested to determine your genetic make-up with the potential that one day a therapy may be available for your particular IRD, please complete the expression of interest form (if you haven't already done so) and send it to the project team in Western Australia.

If you have not received the special mail out containing the Expression of Interest form, please contact the office, on 03 9650 5088, and this information will be sent to you as soon as practicable.



Media Release

Sir Charles Gairdner Hospital, Nedlands, WA 6009
Telephone (08) 9346 3863/ 2404

8 December 2008

Australian first to help researchers seeking eye disease cure

A new national patient register developed at Sir Charles Gairdner Hospital is set to put Australian researchers at a major advantage in their search for a cure for Inherited Retinal Disease (IRD), which causes blindness or severe visual impairment in 15,000 Australians.

The Department of Medical Technology and Physics at SCGH has run a state register since 1984 with funding from the WA Retinitis Pigmentosa Foundation. Now, with a \$360,000 funding injection from Retina Australia, the register will be expanded nationally providing researchers with access to a much larger pool of patients to select for clinical trials.

The WA register has details of more than 1700 subjects, including demographics and results of ophthalmological investigations. DNA samples have also been collected from 800 of those.

President of Retina Australia Graeme Banks said he expected that by 2012 the national IRD Register would include DNA samples from more than 3000 subjects stored in freezers, awaiting genetic analysis.

“The DNA and supporting clinical information will be made available to approved clinicians and scientists looking to identify genetic mutations causing IRD, develop gene therapies or conduct clinical trials,” said Mr Banks.

“It’s a great development that hopefully will lead to breakthroughs in preventing IRDs, slowing their development and ideally identifying a cure.”

“IRDs affect people of all ages and cause varying degrees of visual disturbances that greatly affect the ability of patients to engage in the activities of daily life that many healthy Australians take for granted.”

Principal Medical Physicist at SCGH John de Roach said studies of DNA from the Western Australian register had already identified several genetic defects that were responsible for IRD in 80 individuals.

“Those patients will soon be offered genetic counselling. It is possible that some may become suitable for gene therapy or a clinical trial in the future, as human clinical trials have only commenced for IRD sufferers in the last 12 months,” said Dr De Roach. “These patients and those identified by this project in the future are optimally placed to take advantage of the rapidly developing advances in IRD therapies.”

“At all times we maintain the patients’ anonymity and only if and when they have agreed to participate in a therapy or trial do we provide their information to researchers,” he said.

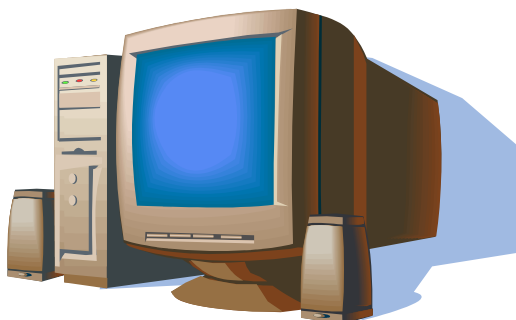
A complete list of DNA samples, associated clinical diagnoses, and any identified causal genetic mutations is updated every 3 months on a public website.

Low Cost PCs for Centrelink Customers

A partnership between Work Ventures and Centrelink now enables Centrelink customers to purchase their own professionally refurbished and internet ready Personal Computers (PCs). All Centrelink concession card holders are eligible to purchase the Pentium 4 PC pack for \$250 plus delivery.

Please note this pack does not include adaptive technology.

Contact your local Centrelink office or phone Work Ventures on 1800 112 205.



2009 Retina Australia Congress

When? Friday 23 October – Sunday 25 October
Where? Royal on the Park Hotel, Brisbane

This year's Congress is shaping up to be yet another fabulous event, promising an extremely informative and enjoyable program. Arrangements are progressing well, and the program will cater for a wide range of interests. The following presenters are included in the program:

- Professor Elizabeth Rakoczy (University of WA and Lions Eye Institute) who researches in the area of genetic treatments for retinal eye diseases.
- Associate Professor Nigel Lovell (University of NSW) who will speak about vision prostheses.
- Dr Erica Fletcher (University of Melbourne) who researches the possibilities of pharmaceutical treatments.
- Dr Monica Acosta (University of Auckland – an associate of Professor Michael Kalloniatis) who conducts research into the causes of retinal eye disease.
- Dr John de Roach (Principal Medical Physicist, Sir Charles Gairdner Hospital, WA) who will speak about the Australian Inherited Retinal Disease Register and DNA Bank.
- Tina Lamey (Senior Research Officer working with John De Roach). She will speak about their work studying the genetics of Western Australian Families Affected by Inherited Retinal Disease.

Sessions of a practical nature in the program include:

- issues relevant to young people
- mental health, particularly depression
- a question and answer session with an ophthalmologist

Enjoy Queensland's warm weather and take the opportunity to find out about the latest research developments directly from the researchers, ask questions, participate in useful practical sessions, and last but not least, have fun and socialise!

Watch out for registration forms which will be distributed next month.

Retina International Supports International Rare Disease Day 2009

Leading vision group Retina International supported the second annual International Rare Disease Day which took place on Saturday February 28 2009. The day was celebrated with many diverse events designed to provide information on this important subject across Europe, The United States and Latin America.

The purpose of International Rare Disease Day is to raise awareness of rare diseases and of their impact on the lives of those affected, their families and society as a whole. It is hoped the day reinforces the importance of rare diseases as a public health priority. The event is organised by members of the European Rare Diseases Organisation (EURORDIS) and its international partners.

Retina International is an umbrella group of 33 national societies promoting research and providing information on rare degenerative eye conditions including Retinitis Pigmentosa (RP), Usher Syndrome, Leber's Congenital Amaurosis (LCA) and Stargardt's disease as well as other allied rare retinal dystrophies. One of the key priorities of Retina International is to foster cooperation between international scientific and patient groups to ensure all the elements are in place to facilitate the development of therapies for rare retinal diseases.

President of Retina International Ms. Christina Fasser said, "It is vital that we continue to raise awareness of chronically debilitating diseases that although considered rare, when viewed internationally can affect millions of adults and children. To facilitate therapeutic development for rare disease it is essential for us to work internationally". She added, "2008 saw great results in two gene therapy trials for a rare retinal disease called Lebers Congenital Amourosis (LCA) both in the UK and USA. Here a small group of young patients had some partial sight restored. To take these trials to the next level a greater number of patients with the same condition will need to be indentified. Access by scientists and clinicians to properly funded, well resourced and internationally coordinated patient registries can speed up this process and, it is hoped, will bring about a cure for rare blinding conditions at the earliest opportunity".

In many countries the needs of patients with rare eye conditions are not being met as there are often barriers to accurate diagnosis and where possible appropriate intervention. Retina International is pleased to show its support by encouraging members all over the world to support local activities on International Rare Diseases Day.

Mr. Yann Le Cam CEO of Eurordis said, "As a direct result of the attention we expect Rare Disease Day 2009 will raise, we hope national health care systems improve the availability and quality of diagnosis, treatment and care for rare disease patients throughout Europe and the world".

There is no cure today for the 6000 - 8000 rare diseases, 75% of which affect children.

2009 Research project to be supported by Retina Australia

'The role of purines in photoreceptor death during retinal degeneration'

Investigators: Dr Erica Fletcher; Professor Michael Kalloniatis;
Associate Professor Claire Mitchell

University of Melbourne

Grant: \$40,000

Lay description:

This project will continue our work examining whether substances released from dying photoreceptors cause the death of neighbouring photoreceptors and whether treatments that block the actions of these released substances can prevent the death of photoreceptors. Our previous work has shown that a compound called adenosine-tri-phosphate (ATP) excites photoreceptors. When cells die, they release large amounts of ATP, that initiates death of photoreceptors. Moreover, our preliminary data shows that compounds that block the actions of ATP, slow photoreceptor loss in animal models of Retinitis Pigmentosa. We propose that when photoreceptor die in Retinitis Pigmentosa, they release large amounts of ATP, that excite neighbouring photoreceptors, to the point where these neighbouring cells die. This project has three parts: **first** we will examine the mechanism by which extracellular ATP induces apoptosis of photoreceptors; **secondly** we will evaluate whether blockading the actions of ATP slows photoreceptor loss in two animal models of retinal degeneration (rd/rd mice, light damage). **Thirdly**, we will quantify the levels of ATP in the vitreous during retinal degeneration. We predict that the results of this project will provide the opportunity to establish a new class of drug for the treatment of retinal degeneration.

Changes to the Multi Purpose Taxi Program



The Victorian Government has introduced important changes to the Multi Purpose Taxi Program in Victoria, which includes:

- the release of a \$14 million package over 4 years
- annual trip caps have doubled to a total of \$2180 per year which will have a significant positive impact for people with a disability. People on an M40 card will still have unlimited travel (ie. no cap).
- the trip cap will be increased from \$30 per trip to \$60 per trip.
- the government are releasing 200 new conventional taxi licences in the network to ensure the greater availability of taxis.
- a number of great initiatives have also been implemented for people who are reliant on wheelchair accessible taxis (WATs).

Blind yachtsman Watson shows he's a more than able seaman

Kirk Watson was on a boat, quietly working away on trimming the mainsail. At the end of a great day on the sea for the corporate clients, Watson picked up a harness and attached it to his dog, Tiller. And then it dawned on those on board. "Ah," noted one visitor, "that is why that dog was there." And Watson was thrilled - no one had noticed he was blind.

Watson, 35, is about to embark on his sixth Sydney to Hobart adventure on avid Pescud's boat, Sailors with disAbilities.

His trusty four-legged eyes, often lying quietly in her special corner on boats for short trips, gets to fly down to Hobart so she can enjoy the dockside festivities.

"She loves a good party," Watson said. On cue, Tiller wags her tail.

Watson has grown up on the water around Sydney, and when his vision started to deteriorate at the age of 16 - a result of a disease called Retinitis Pigmentosa - he was shattered at the thought that he would have to give up sailing.

"I thought I would have to give it up, and I certainly didn't think I would get to the level I have and I certainly didn't think I would do a Hobart," Watson said.

He now has just 2 per cent vision - enough to simply differentiate between light and dark. He got Tiller five years ago, just as he embarked on his first Sydney to Hobart race. He can still remember the feeling of accomplishment and wonder, and imprinted the sight he could faintly make out as the boat was coming into Hobart: the Organ Pipes, spectacular dolerite columns up to 125 metres high on the side of Mount Wellington.

"It is an amazing feeling at the start," Watson said. "Obviously, I can't see how close the boats are, but I am just concentrating on what I have to do on the boat, but the atmosphere is electric.

"And then slowly all the spectator boats drop off and then getting to Hobart is just a great feeling. I can still picture in my mind how beautiful it is around Tasman Island and I can picture the Organ Pipes."

Watson said he worked closely with the helmsman on board to establish non-verbal communication. He laughed and said that mainly consisted of a big shove or a knock to the head to get moving.



"Sailing is not all about seeing - when it is three in the morning and pitch black a lot of the other crew look to me because I know where things are," he said.

A keen surfboard rider and volunteer for sailing with disabled children, Watson said sailing with an entire crew with disabilities helped to raise awareness of disabled people.

"But it is also about giving something back to the community," he said.

Source: <http://www.smh.com.au/articles/2008/12/17/1229189710439>

JACQUELIN MAGNAY, *The Canberra Times* - Canberra, ACT, Australia, 18/12/2008.

RESEARCH UPDATE

\$3 million towards gene therapy to treat retinal degenerative diseases

The Foundation Fighting Blindness (FFB) is partnering with the Canadian Institutes of Health Research (CIHR) in a five-year grant that will fund an ambitious research project to develop innovative gene therapies for a number of degenerative retinal diseases.

This is a milestone in the charity's 34 year history. It involves five research teams, four at prominent universities in Canada and one in the U.S. It's also the first time that the CIHR is partnering with a health charity on a grant to develop therapies for degenerative eye diseases. The partnership will provide a much needed boost to eye research, which still receives significantly less funding in Canada than other major disease areas such as diabetes or cancer.

"This is a huge milestone for the Foundation Fighting Blindness and represents a historic shift in the type of vision research being funded," said Sharon Colle, President and CEO, FFB. "We're taking the knowledge generated through decades of research into causes of these diseases, and are now applying it to the development of long awaited therapies."

The five research teams involved will provide various specialized skills required for the success in this research project. The project will be led by Dr. Robert Molday, a molecular and cell biologist from the University of British Columbia. The team's other

experts in gene therapy are Dr. Jim Hu from the University of Toronto, and Dr. Bill Hauswirth from the University of Florida. Dr. Marinko Sarunic of Simon Fraser University will be responsible for the retinal imaging component of the project. As the team's clinician-scientist, Dr. Robert Koenekoop will oversee the visual function testing and the gene analyses, first in animals and then in humans with a variety of retinal degenerations.

The strategy is to replace the defective gene with a "new healthy gene" in specific animal models for retinal degenerative diseases with the aim of slowing photoreceptor loss and partially restoring vision. Success in these animal models would lead to future human clinical trials.

"The application of gene therapy for three retinal degenerative diseases will be investigated: Stargardt Macular Dystrophy, Leber Congenital Amaurosis (LCA), and Retinitis Pigmentosa," Dr. Molday explains. "The recent success in gene therapy for RPE65 has been highly conclusive for LCA; we believe that we can learn from this and advance even more quickly this time."

RPE65 mutations are one cause of Leber Congenital Amaurosis. Three independent research teams have very recently shown that injecting a healthy version of that gene to young adults can partially restore their vision.

Source: Sharon Colle, President and CEO The Foundation Fighting Blindness, Toronto, December 9, 2008

Lottery Grant for Inherited Blindness Research

Patient-led charity RP Fighting Blindness has been awarded a Big Lottery Fund grant to work in collaboration with London's UCL Institute of Ophthalmology to investigate potential treatments for Retinitis Pigmentosa, a blinding genetic disease of the eye.

The charity is working with Professor Mike Cheetham and his team on a new project looking at the potential for advanced drugs, already approved for the treatment of other conditions, to arrest the deterioration of sight in people with RP.

RP affects about 25,000 people across the UK and over 2 million people worldwide. It is caused by flaws in a number of genes which result in the death of photoreceptor cells in the retina. These cells are where seeing starts, as they convert light into signals to the brain.

David Head, Chief Executive at RP Fighting Blindness, said: "We are proud to collaborate with UCL Institute of Ophthalmology and Moorfields Eye Hospital, which together form a world renowned centre for eye research, and we are extremely grateful to the Big Lottery Fund for their support, without which we would simply not have been able to fund the project.

This is a significant grant and will advance our fight against RP considerably. There is currently no cure but there are several very promising threads of research underway that could lead to treatment, of which this is one. It fantastic news for people with RP, who are anxious of course that every possible avenue of research is followed up."

Prof. Cheetham added: The research funded by the Big Lottery Fund aims to build on an exciting breakthrough in potential treatments for Retinitis Pigmentosa, work that was initially funded by RP Fighting Blindness themselves. My team at the UCL Institute of Ophthalmology in London have shown that certain drugs can counteract the negative effects of mutations in rhodopsin, which is one of the most common causes of RP, using a laboratory model. We now aim to continue the pre-clinical development of these drugs and test if these potential treatments can be taken closer to the clinic. If this approach is successful it could soon be translated as one of the first treatments for rhodopsin RP.

The project will run over three years and commence in early 2009.

Reference: British Retina Pigmentosa Society website: www.brps.org.uk, 21 October 2008

Brain Reorganizes To Adjust For Loss Of Vision

A new study from Georgia Tech shows that when patients with Macular Degeneration focus on using another part of their retina to compensate for their loss of central vision, their brain seems to compensate by reorganizing its neural connections. Age-related Macular Degeneration is the leading cause of blindness in the elderly. The study appears in the December edition of the journal Restorative Neurology and Neuroscience.

"Our results show that the patient's behavior may be critical to get the brain to reorganize in response to disease," said Eric Schumacher, assistant professor in Georgia Tech's School of Psychology. "It's not enough to lose input to a brain region for that region to reorganize; the change in the patient's behavior also matters."

In this case, that change of behavior comes when patients with Macular Degeneration, a disease in which damage to the retina causes patients to lose their vision in the center of their visual field, make up for this loss by focusing with other parts of their visual field.

Previous research in this area showed conflicting results. Some studies suggested that the primary visual cortex, the first part of the cortex to receive visual information from the eyes, reorganizes itself, but other studies suggested that this didn't occur. Schumacher and his graduate student, Keith Main, worked with researchers from the Georgia Tech/Emory Wallace H. Coulter Department of Biomedical Engineering and

the Emory Eye Centre. They tested whether the patients' use of other areas outside their central visual field, known as preferred retinal locations, to compensate for their damaged retinas drives, or is related to, this reorganization in the visual cortex.

The researchers presented 13 volunteers with a series of tests designed to visually stimulate their peripheral regions and measure brain activity with functional magnetic resonance imaging. They found that when patients visually stimulated the preferred retinal locations, they increased brain activity in the same parts of the visual cortex that are normally activated when healthy patients focused on objects in their central visual field. They concluded that the brain had reorganized itself.

The parts of the visual cortex that process information from the central visual field in patients with normal vision were reprogrammed to process information from other parts of the eye, parts that macular degeneration patients use instead of their central visual areas.

While there is evidence with other tasks that suggests that the brain can reorganize itself, this is the first study to directly show that this reorganization in patients with retinal disease is related to patient behavior.

The research group is currently studying how long this reorganization takes and whether it can be fostered through low-vision training.

The research was funded in part by a seed grant from the Georgia Tech/Emory Health Systems Institute.

Reference: Science Daily, 21 November 2008

Researchers Hope to Mime 1000 Neurons With High-Res Artificial Retina

Researchers from three major California universities are working on an artificial retina that could give limited sight to people with degenerative diseases of the retina, such as macular degeneration. Such a prosthesis is a more realistic future treatment than stem-cell therapy, gene therapy, or eye transplants, its developers say. The Californian researchers have been treating people using a 60-pixel retina in a clinical trial for two years.

But they are now gunning for a system with a resolution of 1000 pixels, they reported Tuesday at the IEEE (Institute of Electronics and Engineers) International Electron Devices Meeting (IEDM), in San Francisco. And in contrast with systems in trials today, the researchers hope to develop a system that would be completely sealed into the eye, without any external components.

James Weiland, an associate professor of ophthalmology at the University of Southern California's Biomimetic MicroElectronic Systems (BMES) Engineering Research Center, reported on an experimental system that includes a 1000-pixel test chip. He expects to have the high-res retina at a point where they can begin clinical trials in about five years.

In the artificial retina, a camera mounted on glasses outside the eye sends the visual signals to two RF coils inside the front half of the eye. An electronics module inside the eye's vitreous humor-the gelatinous saline sac that fills the space between the lens of the eye and the retina at the back of the eye-translates the RF signals into voltages for use in the high-res retina chip. Lying against the retina is a grid of 1000 electrodes on a flexible substrate; these electrodes apply voltage signals to the retina, which interprets them as photons. The rest of the visual process takes place as usual, and the system mimics relatively normal vision.

The group, which includes researchers from the BMES center, the California Institute of Technology, in Pasadena, and the University of California, Santa Cruz, which developed earlier prototypes in collaboration with Second Sight Medical Products. The first was the Argus 16, with 16 electrodes; the next, Argus II, has 60. Both have been in clinical trials. The Argus II implant enabled blind clinical-test subjects to follow a straight line for about 6 meters without deviating from the path. But the key to a medical device's ability to grant true independence is whether it allows the person to identify faces or read. Artificial-eye researchers estimate that such tasks will require between 600 and 1000 electrodes.

Ideally, that artificial retina would be contained entirely within a person's eyeball. In order to create a fully self-contained high-resolution system, the team must consider many different pieces: a parylene coating to protect the prosthesis from the corrosive effects of being inside the body for 60 years or more, a flexible substrate that can conform to the idiosyncracies of different individuals' retinal curves, and, most important, wireless power.

Instead of batteries, the device uses inductive coils that pick up energy transmitted from outside the body. The researchers are also relying on insights from MEMS fabrication: the implant coils, interconnects, and 1000 electrodes are formed during a single parylene micromachining process.

"This is a really breathtaking system," says MIT electrical engineering professor Jesus del Alamo, who organized the panel at IEDM where Weiland discussed the group's research. "They have every piece of the system in place-they have even designed their own software." But there is more work to be done. "You need to get everything into the eye," says Jamal Deen, a professor of electrical and computer engineering at McMaster University, in Ontario, "including the camera."

So far, the camera, image-processing hardware, power amplifier, and data modulator are external, but Weiland hopes to implant even the camera part of the system by fixing it to the lens of the eye. His collaborators at USC are working on miniaturizing the camera system so that it can be placed onto the lens in a routine surgical procedure similar to cataract surgery. "If we can make a camera the size of the lens, we can implant it there," he says. "But again, the challenge is making a self-contained camera without a larger control circuit."

He cautions that it will take several years to put the whole system together and start clinical trials. But those trials will lean heavily on what is learned from trials of the implant being tested today. So potential patients should not wait for the new chip. "The 1000-channel device is likely more than five years away from even starting clinical testing," says Weiland. "In the meantime, our 60-channel device has been in clinical trials for over two years, and sometimes we run into difficulty recruiting for the trial because some prospective participants are aware of the research efforts on higher-channel-count devices."

Source: Sally Adee, IEEE Spectrum, 19 December 2008

Progression of Retinal Disease Linked To Cell Starvation

Rods and cones coexist peacefully in healthy retinas. Both types of cells occupy the same layer of tissue and send signals when they detect light, which is the first step in vision. The incurable eye disease Retinitis Pigmentosa, however, reveals a codependent relationship between the two that can be destructive. When flawed rods begin to die, otherwise normal cones follow them to the grave, leading to blindness. A new study might explain why.

Data published online in Nature Neuroscience Dec. 7 suggest the cones are starving to death. As rods disappear, the structure of the retina breaks down. This might disrupt the connections between the cones and their source of nutrients.

"This is the first study linking cone death in Retinitis Pigmentosa to a metabolic problem that suggests starvation," says senior author Constance Cepko, an HMS professor and investigator with Howard Hughes Medical Institute. "If we can find a way to supply nutrients to the cones, we might be able to preserve daylight vision in patients."

Active in bright light, cones allow us to perceive color and fine details. Conversely, rods allow us to see in dim light. The untrained eye cannot distinguish between the two types of cells, which grow side-by-side. Both rods and cones have a protrusion that has many membranous discs, resembling a stack of cookies. A cone stack is half the height of a rod stack. Stacks emanating from both types of cells get clustered together, like Oreos on a plate. The entire plate gets covered in "plastic," with the flexible plastic

reaching down to touch each stack. In the eye, this plastic consists of a giant retinal pigment (RPE) cell, which supplies nutrients to the rods and cones on its plate.

With this structure in mind, researchers have proposed a variety of hypotheses to explain the loss of cones in patients with mutations in rod-specific genes. For example, some teams have suggested that rods produce a chemical cones need to survive. But the data didn't quite fit the proposed models.

Cekpo's team took a fresh approach to the problem. Postdoctoral researcher Claudio Punzo gathered four strains of mice, each with a different rod-specific mutation and a different rate of disease progression. He discovered an interesting pattern. Cone death always began after the major phase of rod death.

Punzo analyzed gene expression before and after this point in each strain. During the cone death phase, 230 genes were always expressed at higher levels. Sleuthing revealed that 34.9 percent of those play a role in cellular metabolism, including 12 genes in the insulin/mTOR pathway.

mTOR serves as a signaling hub, gathering information about the environment and helping the cell to decide whether it has enough nutrients to make new proteins. Punzo now had a lead. Further experiments suggested the cones weren't getting enough glucose. Not only did they express high levels of a protein that allows the cell to take up more glucose, but the cones survived longer when Punzo tricked them into thinking they had enough glucose by injecting the mice with insulin.

"Apparently, the cones caught onto our trick," says Punzo. "After surviving longer than usual, they started to die in droves."

Cepko and Punzo say the new hypothesis makes sense. Rods outnumber cones by more than 20 to 1. The RPE cells sag when too many rods disappear, as the plastic over that plate of Oreo cookies droops when too many stacks are missing. The structural change likely disturbs the contacts between RPE cells and cones, impeding the flow of nutrients to the cones.

"This points us in a new direction," says Cepko. "We're currently exploring ways to boost nutrient levels in the cones. Perhaps someday we can help Retinitis Pigmentosa patients maintain their daylight vision for at least a bit longer than they otherwise would."

This research is supported by the National Institutes of Health, the Macular Research Foundation, The Foundation for Retinal Research, Merck and the European Molecular Biology Organization.

Source: Science Daily, 7 December 2008

Question Time

with Jessica Zammit

In this edition, member Jessica Zammit has kindly agreed to volunteer for Question Time. Please contact Rick through the office to volunteer your answers for future editions.



1. What's your earliest memory?

Playing doctors and nurses with my male best friend in kindergarten and photo day where I had to wear my favourite pink and white pocka dot dress (it was the 80s after all) with an ugly skivvy underneath because it was too cold outside. See even back then I was fashion conscious! *grin*

2. What's your idea of a good time?

Going somewhere new and trying something different – be it a new restaurant or exploring a new city. It's also spending time with good friends with a great cocktail and even better conversation. A dance or two doesn't hurt either...

3. What's your ideal holiday destination?

Europe – there is something really quite beautiful and each country has

so much to offer. In 2005 I travelled around Europe briefly but also spent some time in Sri Lanka which was amazing, with its beautiful mix of jungles, desert, pristine coastlines and endless views. If you have the opportunity, it is definitely worth visiting.

4. Who inspires you?

There is more than one person who inspires me. It's easy to mention a person that everyone knows but I'm often inspired by the stories I hear from everyday people who have faced adversity but have come out stronger. I am also inspired by people who challenge the status quo – the Barrack Obamas of the world who become president as well as those who have simpler ambitions and the optimism to reach their goals.

5. What makes you angry?

People's ignorance in regards to disability – there is so much that people with a disability can and do achieve which is often overshadowed.

6. What's the hardest thing you've ever done?

Breakups.

7. What's the best thing you've ever done?

Travel independently. It forces you to get outside of your comfort zone and meet new people. Whilst I love travelling with others, I'd love the opportunity to travel around parts of the world independently to really put myself out there.

8. What do you like about Retina Australia (Vic)?

I like the fact that Retina is small in size. This might be an unusual compliment, but it means that you can call up and almost always speak with an office member who greets you warmly and puts themselves out there to provide support if you need it. This is pretty rare in the disability sector particularly with larger service providers.

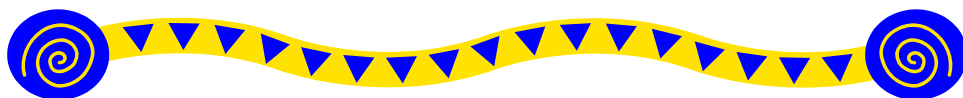
9. If you could change one thing about the world, what would it be?

Collective consciousness – increasing awareness of what is

happening in people's community and around the world rather than people focusing on their own insular, individual concerns. Positive change would be so much more achievable.

10. What's the most important thing you've learnt about life?

To live it. It's so easy to fall into the trap of the day to day work grind and forget to enjoy the little things. I remember reading a story of a woman who was diagnosed with breast cancer. She told of a special bottle of perfume that she adored and because she adored it she only used on special occasions. When she found out she was dying, she started using the perfume everyday and told her friends not to keep the special things for special occasions, but to enjoy the things and people you love everyday, because everyday is special. This really stuck with me and is a philosophy I try to fulfill.



LAST WORD

The greatest discovery of my generation is that a human being can alter his life by altering his attitudes of mind.

To improve the golden moment of opportunity, and catch the good that is within our reach, is the great art of life.

WILLIAM JAMES (1842-1910)

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