Thumbs UP

MND / ALS
ASSOCIATION OF SOUTH AFRICA

The Newsletter of the
Motor Neurone Disease /
Amyotrophic Lateral Sclerosis
Association of South Africa

September 2010

Jozanne at the launch of her Book, "I Choose Everything" with her husband Dave, son Luke and daughter Nicole. (See story inside)

The Mission Statement of the MND Assoc. of S.A. is
"To provide and promote the best possible support for people living with
Motor Neurone Disease, their families and carers and to raise public awareness."

MNDA of South Africa is a member of the International Alliance

PATRON	NATIONAL CHAIRPERSON	VICE CHAIRPERSON
Aviva Pelham (M. Mus.)	Dr. Franclo Henning	Sheila Kendal

SECRETARY
Rina Myburgh

ACKNOWLEDGEMENTS
John Hall for producing the design and DTP (desk top publishing) work.
Logo Print (Maitland) for the printing of our magazine

It is almost unbelievable that the year has gone by so quickly. With all the excitement of the 2010 FIFA World Cup now a splendid memory (and looking forward 2011!), we have the festive season winking at us! And as you know, that means it is again time for sending Christmas cards to family and friends – so, please, get your cards from us – the order forms are enclosed.

Mandy Snyman, a patient from Port Elizabeth, who put together that magnificent concert in August 2008, has compiled a **CD of Christmas Carols** "Let's put back the feeling in Christmas" which she is offering to our readers at R75 each, in order to raise funds for our Association and Hospice. This would make an ideal **Christmas gift** and we urge you to contact her on 041 365 3615 or her sister Sheryl on 082 894 4524 to place your order.

NOTICE IS HEREBY GIVEN OF
MNDA OF SA
ANNUAL GENERAL MEETING
TO BE HELD ON **FRIDAY 8 OCTOBER 2010** AT 10 AM
AT PINELANDS
Parking is available at Howard Centre parking area.

PLEASE - advise our office on 021 531 6130 *(or e-mail: mndaofsa@global.co.za)*
of your attendance by latest ***Monday 4 October*** *to facilitate seating and catering arrangements*

Donations and Thank You's

We once again want to thank all our regular donors for their loyal support during the year as well as all other donations from various sources. Your financial support is so much appreciated, relying as we do on voluntary donations, membership fees and our own fundraising efforts for our income.

We need to say a big thank you to some other rather special people who have given their time to so generously assist us with various tasks during the past year:

John Hall *for doing the design and layout of Thumbs Up,*
Logo *Print for printing Thumbs Up ,*
Hilda Hunter *for labelling and filling the envelopes for posting,*
Elsabe Burger *for taking charge of Christmas Card sales*
Peter Rosmaren *for checking our accounts before it goes to the auditors, and*
FMG Inc *who conducts our audit and compiles our financial statements*
(If someone's name has been omitted, it is by accident and not intent)

---

21 June each year is ALS/MND Global Awareness Day
People who care make a world of difference
A tracking survey in the UK has shown that awareness has increased in the last five years, despite the fact that only half of people were told of the Association at the time of their diagnosis

A patient's feelings -

When I was first diagnosed with MND my whole world collapsed around me. The only people I could face telling were the two people closest to me, my wife and my daughter. After a lot of sleepless nights, we realised we needed help and support from people who were going through, or have been through, what we were going through.
So I summoned up the courage to ring MND Connect*. Suddenly we felt we were not alone. Things started to feel better. It was suggested that we go to support group meetings. We felt very uneasy about this at first, were not sure whether we were ready to meet people living with MND who were in more advanced stages than I was.
We decided we would give it a go. On the morning of our first meeting we were all very nervous and not feeling at all sure, we very nearly decided not to go at all but we summoned the energy and went along. We were met very warmly and taken inside and introduced . We were well looked after with a nice cup of tea and home made cake !
How glad we were that we made the effort to go along. We have met so many good people, people that are in the same boat as us and know how it feels, people we can call our friends.
(Extract from Thumbprint Magazine (UK)*

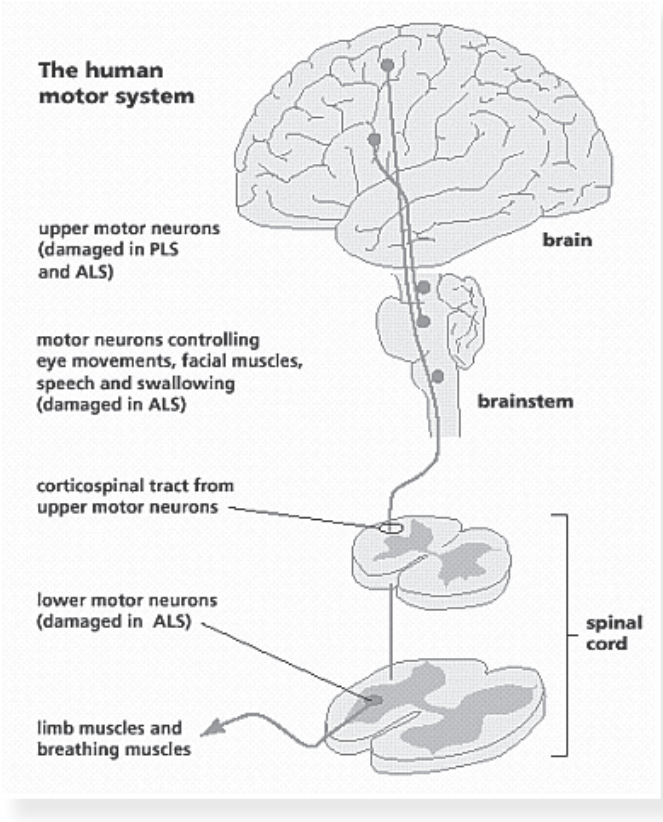
FROM THE CHAIR

From the Chairman, Dr Franclo Henning, Neurologist, FC Neurol (SA)

Dear Friends

Some of you may have noticed that the logo at the top of this newsletter has changed. Instead of MNDA of South Africa, we have decided to change our name to MND/ ALS Association of South Africa. This change came about when we realized that a large portion of the public, patients and health care workers are not aware that these two terms are used interchangeably (sometimes incorrectly). I will briefly try to explain the meaning of each of these terms, as well as a few other relevant terms.

The term motor neuron disease (MND) refers to a group of disorders affecting motor neurons, the nerve cells that are involved in the control of "motor function", i.e. the contraction of muscles. There are 2 populations of motor neurons – upper motor neurons (UMNs) and lower motor neurons (LMNs). Upper motor neurons start in the brain and end in the spinal cord, while lower motor neurons start in the spinal cord (called the anterior horn cell) and end at the junction with the muscles they supply. Have a look at the illustration below to understand it better.



Motor neuron diseases can therefore affect either upper motor neurons, lower motor neurons, or both. Based on which population of motor neurons is affected, 3 different disorders can be distinguished:

Amyotrophic Lateral Sclerosis (ALS) - affects UMNs and LMNs

Primary Lateral Sclerosis (PLS) -affects UMNs only

Progressive Muscular Atrophy (PMA) - affects LMNs only

Of these three, ALS is by far the most common, affecting about 90% of patients with MND, which is partly why many people use the terms MND and ALS interchangeably. People with ALS typically develop weakness, stiffness (spasticity), swallowing and speech difficulties, fasciculations (muscle twitching) and atrophy of their muscles. This disease is often (but not always) rapidly progressive over 3 to 5 years. If the disease starts with speech and swallowing abnormalities (bulbar onset ALS), disease progression is often more rapid.

PLS typically causes leg weakness and spasticity, and may later involve the arms and swallowing / speech (milder than in ALS). PLS is a very slowly progressive condition and patients often die of other conditions unrelated to PLS.

PMA consists of a few subcategories, which becomes complicated, but can mainly be divided into generalized (affects most muscle groups of the body) and segmental (affects limited groups of muscles). Generalized PMA may appear similar to ALS, but does not cause spasticity and other upper motor neuron problems. The course also seems to be very similar to ALS. Segmental PMA causes mainly weakness, fasciculations and atrophy of certain muscle groups, and is also very slowly progressive.

Based on this explanation you can certainly ask what the reason for the name change is. The term MND does appear to be appropriate, as it includes all the disorders our association strives to care for. However, it became quite clear to us that some patients diagnosed with ALS, their families, and even potential benefactors are not aware that ALS is a form of MND, and hence never come in contact with the MND Association of SA. In fact, when a play based on a well known book about a man with ALS was recently staged locally, the organizers could find no reference to a local organization caring for patients with ALS, and instead donated the proceeds to a different organization. By incorporating "ALS" into our name, we hope to make our association more accessible and visible to patients, their family and friends and the general public.

MAIL BAG

Gail Collins of Barberton, Mpumalanga, wrote to us:

Our beloved husband, father and best friend passed away peacefully on 3rd June 2010. Brian was diagnosed in March 2008 by a neurologist in Pretoria and then we started our journey of the unknown.

Thanks to Liz Keth, who stays about 400 km's away from us, for her valuable advice and assistance.

We tried to do the things together that we wanted to do once we retired. We travelled to Australia to watch a five day cricket match. We thoroughly enjoyed visiting my family in Melbourne

and then went to Sydney to watch the 2009 New Year test.

September 2009 we flew to Kenya to visit Brian's brother and visited a game park and had wonderful sightings of all game and birds. During our stay at a mountain lodge we were given a room on the 1st floor because the dining room would be closer for us. There was no lift, so 4 porters carried Brian up in his wheelchair, to our room. That was an experience all on its own.

We visited Kruger a few times as they have perfect "mobility challenged" chalets - Highly recommended.

In March Brian was admitted to hospital to insert a feeding tube. He was struggling to swallow. The specialist physician and the anaesthetist were not happy with the state of Brian's lungs. They decided to sedate Brian and do a local anaesthetic. I was then able to feed Brian with Ensure and whatever medication, liquids and vitamins he needed.

About 1 month before Brian passed away our Deaconess Sheila, from our church, came to visit and we had communion together. Brian was adamant that he wanted this, so we crushed the wafer, mixed it with the grape juice and fed it to him via the tube. This experience was shared at the funeral and Sheila expressed Brian's strong faith and commitment to our Lord and Saviour Jesus Christ.

I strongly urge each and everyone to make peace with your disease and seek a professional Christian counsellor.

Thank you to MNDA of SA for your tireless work.
Regards
Gail, Stephen & Michael Collins

Sharon Smith the daughter of John Mansfield, a MND patient from the UK who passed away recently, placed this poem, written by her late father about how he felt about his wheelchair, in the *MNDA (UK) Thumb Print Winter 2010 issue:*

The Wheelchair
Two large wheels with handgrips, two smaller ones too
Two handles, a seat, and a frame coloured blue
It waits in the hallway whilst I'm in my bed
Aware of its duty – to act as my legs

Arms ever open, inviting me in
I want to resist but I know I can't win
Up, turn and lower has me safely enthroned
Am I stuck with this illness or has fate only loaned ?

Get myself settled, there's a long day ahead
Wheels will replace these legs of lead
I wait for the movement that just will not come
So hands to the wheels – push and run, push and run

Friendly hands offer help, I'll accept if I must
It's away to say thank you to those I trust
The wheelchair – perhaps I should thank it and call it a friend
But I'll also resent it – right up to the end.

Jozanne's Book Launch

Jozanne Moss, who was diagnosed five years ago with motor neurone disease (MND), co-authored a book with Michael Wenham, a British Anglican priest who has primary lateral sclerosis, to tell their incredible and touching stories of living with these degenerative diseases. The two authors were introduced via the internet in 2009.
A sparkling Jozanne launched her much anticipated book, "I Choose Everything" both at home and at the Baptist Church last week with her family - husband Dave and children Luke (12), Nicole (10) and almost 150 people.
Luke and Nicole proudly assisted in selling and 'signing' while Dave explained the title of the book, so seemly suitable because Jozanne lives life to the fullest. Always when someone asks her what she likes about Dave, her answer would be, 'I choose everything'.
Luke, a keen classical guitarist and Nicole, an upcoming pianist surprised their mom with their beautiful music. Friend and pastor, Neill Tucker said to Luke and Nicole: "Look around you - this is your new family in Christ."

Southern Cape Support Group
*From left, seated****: Patients Leon Groenewald from Oudtshoorn, Jacques Rossouw of Wilderness with Jozanne Moss and standing, Dilys Jones (Southern Cape Support Group Facilitator) and Sheila Kendal (MND Consultant and vice Chairlady) right.***

A second book launch was held at their home with Jozanne and a concourse of friends, family and those who are supporting her through her illness.
The book @ R120 can be obtained from Dave on 079 797 9049

News from Liz Keth, MND Consultant for the Gauteng area

Photos from our 23 June 2010 support group meeting which was held at a family restaurant in Randpark Ridge. Monthly meetings are held on the last Wednesday of every month where patients share with one another over a cup of coffee, or where we have a guest speaker talk on various aspects of common interest.
Contact Liz on 011 701 5001 or 082 878 3716 for any information regarding the meetings.

NEW INFORMATION OFFERS SUPPORT

Cognitive impairment and its impact for people living with MND:

Initially, MND was thought only to be confined to the motor system which controls movement and it is only recently that there has been an increased understanding of changes in the brain that affect thinking, emotion and behaviour.

Working with people living with MND it is important to increase awareness of cognitive change and more importantly, how to help people understand ways to support them to adjust to the changes.

People can feel a tremendous sense of isolation when cognitive change occurs, often not knowing where to go for help. At diagnosis it isn't unusual to hear people commenting 'at least MND doesn't affect the mind', so people often feel that they are facing a double diagnosis when we begin to talk about cognitive change.

What does 'cognitive change' mean:

Cognition refers to a range of brain functions that include the ability to learn new things, think through and plan activities, concentrate and carry out more complex and demanding tasks. Cognition also includes language abilities such as the capacity to generate words and the ability to interact with and respond appropriately to other people. When we speak of 'cognitive change' we refer to any changes in some or all of these abilities.

Who does it affect:

People with MND appear to fall into one of four groups:

1. Those whose cognitive function is unaffected
2. Those who demonstrate subtle cognitive change and/or behavioural changes
3. Around 5% of people with MND develop a condition called 'fronteotemporal dementia (FTD)
4. Those with FTD who go on to develop motor impair ments and MND diagnosed after Dementia

The degree of cognitive change experienced by people varies enormously, some
people notice only mild symptoms, others find that changes impact on their daily lives to a
greater degree. It is important to distinguish mild changes from a type of MND related
dementia called frontotemporal dementia (FTD). As the name suggests this is a form of
dementia causing severe cognitive and behavioural change.

What are the symptoms of cognitive change ?
They can include: difficulty concentrating perhaps when reading or dealing with bills, learning
to use new equipment such as computers or new activities, becoming easily distracted in
noisy environments and struggling to finish tasks.
Some complain of problems 'multi tasking' such as ironing and watching television at the
same time, people can become easily distracted. Carers, family and friends may be the first
to notice subtle changes, for example the person isn't quite 'themselves' in responding to
social situations appropriately or perhaps language seems a problem in that they forget
names of daily objects or tasks and may struggle to understand complex conversations.
It is also important to note that for some people, cognitive decline can be so subtle that it
may not even be noticed until the person has undergone a detailed assessment.

What do you mean by behavioural changes?
We sometimes hear about symptoms which make us wonder if a person is experiencing
behavioural changes. Such changes can include feeling restless, acting impulsively, eating
lots of sweet foods sand cramming food, becoming fixated on an activity or routine,
becoming indifferent to the distress of a partner or carer.
Occasionally it is the person's career or family member who wants to talk about these
Symptoms in clinic and they may feel that the person isn't actually aware that there is a
problem.

What else could it be?
We need to rule out other causes such as changes in breathing or presence of infection,
which can also affect some aspect of cognition and behaviour.

RESEARCH

(Extract from MNDA (UK) magazine, Thumb Print, Winter 2010 issue:)

Major funding into MND research:

A major new £4.6 million research initiative involving leading MND researchers has been announced by the Wellcome Trust and the Medical Research Council (MRC).

It has long been suspected that similarities exist in the way nerve cells degenerate in different neurological diseases. Several recent discoveries suggest that this is particularly true for MND and a form of dementia called fronto-temporal dementia (FTD), which both have abnormal deposits of the RNA processing proteins TDP-43 and FUS.

Building on his recent discovery of TDP-43 and FUS gene mutations in MND patients, Prof Chris Shaw, based at King's College London, is leading an international consortium of researchers to develop a series of new disease models that will mimic key features of MND and FTD in the laboratory.

"Recent discoveries have placed these RNA processing proteins 'centre-stage' in both MND and FTD', explains Prof Shaw. "RNA processing is vital to cellular function and survival, as it governs the manufacture of all proteins which are the building blocks of all cells. As our nerve cells are built to last us a lifetime, any disruption of this process will have damaging long-term consequences.

"We are extremely pleased to have received this grant. The new disease models we develop will help us to understand the biochemical events that initiate the degeneration of nerve cells in these disorders and will be used to accelerate drug discovery."

Prof Colin Blakemore, one of the UK's most respected scientists, welcomed the announcement:
"This major grant from the MRC and the Wellcome Trust is another indication of the recognition of the importance and broader relevance of research on MND.

"The discovery of a number of genetic mutations that underlie the rare inherited forms of MND could help us to understand common, non-inherited forms of MND but also other degenerative diseases of the nervous system. It is wonderful to see this concentration of funding aimed at understanding the origins of these terrible diseases".

Dr Brian Dickie, director of research development, added: "This is a wonderful example of scientists from different disciplines coming together to work on a common problem. The collaborative thinking of the researchers has been made possible by the collaborative funding from the UK's major medical research bodies. This research could make major inroads into understanding MND."

The Wellcome Trust funds innovative biomedical research, in the UK and internationally, spending over £600 million each year. The MRC invests in world-class scientists to enable them to tackle research into the major health challenges of the 21st century.

Encouraging results from trial

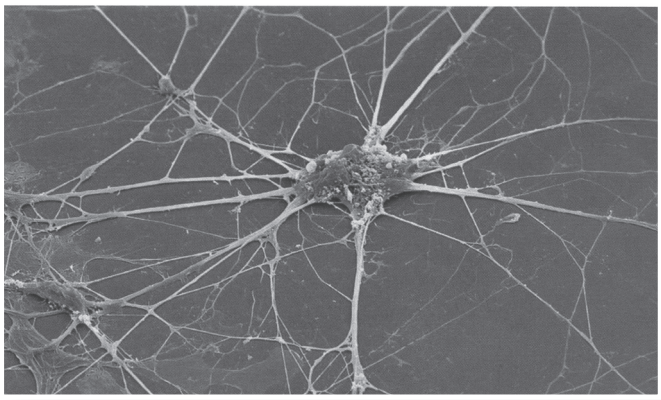
Researchers have presented the encouraging results of a Phase 2 clinical trial looking at the safety and tolerability of a drug called KNS-760704 in ALS patients. ***ALS (Amyotrophic Lateral Sclerosis) is the most common form of MND.***
The two-part Phase 2 trial found that KNS-760704 was safe and well-tolerated in ALS patients for up to nine months. The trial only took place for nine months. The trial results also showed trends suggesting the potential for reducing the rate of decline in the functional capacity of patients for example, how far people can walk unaided, lifting objects, dexterity in eating, and survival.

The researchers emphasised that KNS-760704 is still in the early days of development for patients and that further testing in a large, longer-term, carefully monitored Phase 3 trial is needed to establish the necessary evidence that the drug is both safe and effective for ALS patients.

Dr Belinda Cupid, research manager explains: "Phase 2 clinical trials determine the size of the dose, the timing of the dose and how the drug is to be taken for the next phase of testing. Although Phase 2 testing provides some indication of a drug's ability to treat a disease, the number of patients involved at this stage is much too small for the findings to be relied upon.

"Phase 3 clinical trials are important as they aim to show whether or not the drug actually has a beneficial effect on patients. This stage of testing will usually involve hundreds of patients which is enough to allow a reliable assessment of the drug's effectiveness. Phase 3 results will determine whether or not a drug is to be approved to treat a disease".

Phase 3 testing of KNS-760704 in ALS may begin in mid-2010 at sites in Europe and North America. If you are affected by MND and would like to know more, you can contact Tom Petzinger at Knopp Neurosciences Inc at tom@knoppneurosciences.com



A motor neurone (Steve Gschmeissner)

Harnessing the power of stem cells

(Extract from MNDA (UK) magazine, Thumb Print, Summer 2010 issue:)

In May it was announced that the MND Association is funding its first ever stem cell research programme to help unlock the secrets of MND.
Pulling together world-class expertise from four leading researchers, Prof Siddharthan Chandran, Prof Sir Ian Wilmut, Prof Chris Shaw and Prof Tom Maniatis, the groundbreaking programme will enable scientists to perform detailed laboratory studies on human motor neurones containing known causes of MND.
The ability to grow and programme human motor neurones in the laboratory has been a holy grail for MND researchers for many years. Recent advances in stem cell research mean it's now possible to generate human motor neurones from donor skin cells in order to study the processes at work in health and disease.

The £800,000 three year international research programme will develop and characterise human brain cells, derived from the skin cells of MND patients with the hereditary TDP-43 form of the disease and also from 'control' donors who do not have MND and carry the normal TDP-43 gene.

Why TDP-43?
The TDP-43 gene appears to be a direct cause of MND in around 1% of cases but the protein that the gene produces is found in up to 90% of MND cases. This discovery has been described as 'a seismic shift' in understanding the disease, as it points to TDP-43 playing a pivotal role in many forms of MND. The TDP-43 protein has also been implicated in other conditions – in particular some forms of dementia – so it may prove to play a contributory role in a wider number of neurodegenerative diseases.

The research
The skin cells are initially 'reprogrammed' to generate induced pluripotent stem cells (iPS cells) which are very similar to stem cells derived from human embryos. The iPS cells can then be modified to turn into either of the two main cell types known to be involved in the disease: the motor neurones which degenerate in MND and other vital support cells called astrocytes.
Dr Brian Dickie, director of research development, says: "Although it's the motor neurones that die in MND, we know the disease process is not solely restricted to these cells.
The support cells that normally play a role in nurturing motor neurones can inadvertently cause damage, and it's through this mechanism scientists believe the disease spreads from one part of the brain and spine to the next – a little like the wind fanning the flames of a forest fire."
The research programme will address a fundamental question of whether the support cells from healthy or TDP-43 mutant gene carrying patients are injurious or protective to motor neurones. In addition, experiments growing motor neurones and support cells from different donors in the same dish (a process called 'co-culture') will address whether this disease spread seen in the human condition can be reproduced in the laboratory.
Prof Chandran, the programme's principal investigator, will be working with Prof Wilmut to refine and optimise the reprogramming procedure to ensure as many healthy cells as possible can be generated.
Studies to examine the general health and structure of the different cell types when interacting with each other will be carried out at the University of Edinburgh and by Prof Shaw at King's. Prof Tom Maniatis in New York will work on the programme's third stage by identifying 'gene expression signatures' associated with MND. This means showing which genes within the cells are being switched on and off at various times as the disease evolves.

We welcome your ideas – *THUMBS UP* is your voice –
So if you would like to Contribute to the next issue – Please write to us !

Prof Shaw explains: "Pharmaceutical companies have drug libraries containing millions of potentially useful compounds and yet we can only test a handful of these in clinical trials every year. A key goal of this research is to grow motor neurones from patient skin cells in order to identify which drugs can slow or reverse the disease process. It will be a huge challenge, but if we are successful it will be a great step forward in advancing drug discovery.
This is a highly promising field of research to help increase our understanding of this disease", Brian Dickie continues. "The outcomes from our stem cell research programme will have a powerful impact in shaping the future of MND research and enhancing future international research collaboration".

A few clever "daffinitions"

ADULT - A person who has stopped growing at both ends and is now growing in the middle
DUST - Mud with the juice squeezed out
YAWN - An honest opinion openly expressed
RAISIN - A grape with sunburn
SECRET - Something you tell to one person and a time
WRINKLES - Something other people have, similar to our character lines
INFLATION - Cutting money in half without damaging the paper
(Something we can all relate to !)

Disabled Travel

New Additions

-Epic Guesthouse in Noordhoek, Cape Town
-Mountain Zebra National Park near Cradock
-Eurentia Apartment in Jeffrey's Bay
-Addo National Park (Matyholweni Camp) has a great new picnic area with accessible facilities as well as a new educationl centre. Unfortunately the shop is still not accessible and the new lapa with gravel stones as floor cover is very difficult to access.

HELPING THROUGH YOUR WILL

Your Will can be a convenient vehicle for making a charitable gift of a lasting value. Please consider MND Association as a living memorial for a loved one. Many people support the work of the MND Association of South Africa through bequests from their Estates.

A great place for a break is the ***holiday town of Hartenbosch*** near Mossel Bay. The *ATKV indoor pool* has a hoist (pictured here) to transfer into the lovely heated pool with water jets and seating areas in the pool. The armrests of the chair can be removed for transfer.

Items offered for sale by Janet Meyer whose husband Con passed away recently:

A Sandman dou bilevel Cpap machine with built in humidifier. It also has an opus 360 nasal pillow
An electric wheelchair with 4 wheels – speed 12km/hr and 20 km range with headrest
A right foot support Blue Rocker. Carbon fibre. Made in Scandinavia. Size med 7/8
(Above items have only been in use for 6 months.
Janet can be contacted on : 021 887 4395 or 084 580 1298 or email: janetmmeyer@gmail.com

Condolences to Family and Friends of :

Johanna de Kock, Connie Pretorius, Brenda Day-van Heerden, Isabel Davis, Vonnie van Wyk, Ronel Cloete, Brian Collins, Gavin Swanepoel, Eddie Thathiah, Denzil Sonemann, Con Meyer, Melanie Marais, Lesley Birkett, Shemduth Jugnundan, Gert Smit, Michael le Roux, Elwyn Pelser, Roelie Uys, Jakes Adonis, Isaac van Zyl, Colin Cohen, Abdul Arend, Jean Lowe, William Farmer, Gideon Lombaard, Anna (Barbie) Archer, Oswald Schoeman

MOTOR NEURONE DISEASE ASSOCIATION of South Africa
P.O.Box 789, HOWARD PLACE, 7450,
TEL: (021) 531-6130
FAX: (021) 531-6131
E-mail: mndaofsa@global.co.za
JOHANNESBURG
TEL: (011) 701-5001
CELL: 082-878-3716
E-mail: keth@telkomsa.net